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An anatomical and retrospective clinical study of interventricular septal defects

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AN ANATOMICAL AND RETROSPECTIVE CLINICAL STUDY OF INTERVENTRICULAR SEPTAL DEFECTS

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of
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Declaration

I, Mohseena Osman declare that this dissertation is my own work. It is being submitted for the degree of Master of Science in Medicine in the University of the Witwatersrand, Johannesburg. It has not been submitted before for any degree or examination at this or any other University.

M. Osman

12th
---------- Day of October, 2012

In memory of my father
Ahmed Rasheed Osman
1952-2010

Abstract

A ventricular septal defect occurring on its own is a congenital defect of the interventricular septum of the heart causing varying degrees of increased pulmonary blood flow and associated clinical symptoms. It may also occur in association with obstruction in the right ventricle resulting in diminished pulmonary blood flow. Very little is documented about the incidence of ventricular septal defects in South African children.

The aim of this study was to briefly review the embryology and consider the normal anatomy of the interventricular septum. In addition, the clinical and surgical notes of all children that underwent surgical repair of ventricular septal defects (these included isolated ventricular septal defects, those with multiple ventricular septal defects, as well as those associated with tetralogy of Fallot and double chambered right ventricle) referred to the paediatric cardiothoracic unit at the Charlotte Maxeke Academic Hospital, from the Paediatric Cardiology units at the Charlotte Maxeke Academic Hospital, Chris Hani Baragwanath Academic Hospital and the Rahima Moosa Hospitals between 2001 and 2004, were analysed for the position, number of and size of ventricular septal defects.

For this purpose, 11 cadaveric neonatal hearts were dissected while seven post-mortem specimens of the heart, four with isolated ventricular septal defects and three with tetralogy of Fallot were analysed. In addition, 50 cases of isolated ventricular septal defects, 42 cases with tetralogy of Fallot and eight cases with double chambered right ventricle were retrospectively reviewed.

The membranous and the muscular septum made up the largest components of the interventricular septum in the normal neonatal hearts. A perimembranous ventricular septal defect was the most common type of defect diagnosed in these patients (the majority of the

patients were Black children), 78% of cases with a single isolated ventricular septal defect, 90% in patients with tetralogy of Fallot, and 75% in the group with double chambered right ventricle. The average sizes of all perimembranous defects (from the echocardiogram) were classified as small, moderate or large. An additional interesting finding was prolapse of the right coronary cusp of the aortic valve into the isolated ventricular septal defect and occurred in 22% (11 out of 50) of patients. Aortic regurgitation occurred in association with prolapse of the right coronary cusp in 54.5% (5 out of 11) of cases. This prevalence in black patients is much higher than has been documented in White or Japanese children.

Non-compaction of the left ventricle was seen in one post-mortem specimen in association with a perimembranous ventricular septal defect and dilated cardiomyopathy. This recently described abnormality is alluded to in the discussion.

Attention is drawn to the abnormality known as double chambered right ventricle, where the size of the ventricular septal defect is variable and where the site of obstruction in the right ventricle is caused by muscle bundles lower than that seen in tetralogy of Fallot.

Knowledge of ventricular septal defects found in South African children will help in the assessment and care of these patients with one of the most common congenital cardiac malformations.

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It is with great pride, that I am able to present this research. It would however not have been possible without the assistance and commitment of some dear friends and colleagues. It is with the utmost appreciation that I thank Professor Beverley Kramer for agreeing to supervise this project and also for her assistance, guidance, encouragement and support throughout its writing. For his time and patience, long dedicated hours , in depth tutorials on this subject, as well as for the use of his postmortem specimens, without which this dissertation would not have been possible, a very special thank you to Professor Solomon Levin.

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1. INTRODUCTION

A ventricular septal defect is a congenital defect of the interventricular septum of the heart causing varying degrees of increased pulmonary blood flow and clinical symptoms (Krabill, 2004). A ventricular septal defect is the most likely cause for a congenital heart abnormality which is acyanotic and accounts for approximately 20% of all congenital cardiovascular malformations (Krabill, 2004). In the case of a cyanotic child with congenital heart disease, it is tetralogy of Fallot (which includes a ventricular septal defect) which is the most likely lesion (Shinebourne and Anderson, 2002). The tetralogy of Fallot accounts for 4-10% of congenital heart disease.

The haemodynamics of a ventricular septal defect is that of a left to right shunt with diastolic overload to the left ventricle, pulmonary plethora, pulmonary hypertension and biventricular hypertrophy (Krabill, 2004). If the defect is large, the haemodynamic changes lead to cardiac failure and poor physical growth. Therefore, open heart surgery is necessary to close the ventricular septal defect. The haemodynamics of tetralogy of Fallot is that of a right to left shunt from the right ventricle to the left ventricle with desaturation (cyanosis) on the systemic side of the circulation (Shinebourne and Anderson, 2002). These haemodynamic changes are due to the combination of a large ventricular septal defect and right ventricular outflow tract obstruction.

Other features that may be associated with a ventricular septal defect include prolapse of one of the aortic leaflets of the aorta (Ando *et al*., 2003). This in turn may result in aortic regurgitation. Another anatomical aberration which has been recently recognised in association with

ventricular septal defects is non-compaction of the left ventricular myocardium (Freedom *et al*., 2005). The presence of non-compaction may worsen the prognosis for survival.

In order to understand ventricular septal defects, the development and normal anatomy of the interventricular septum will first be explored and the possible anatomical locations of a ventricular septal defect will be examined.

1.1 Development of the interventricular septum

The cardiovascular system is the first major system to function in the embryo. The primordial heart begins to develop during the middle of the third week post-fertilisation. The cardiovascular system is derived mainly from splanchnic mesoderm, which forms the primordium of the heart, as well as from neural crest cells (Moore and Persaud, 2003). In the cranial part of the intra-embryonic mesoderm, a condensation, which is the cardiogenic plate, occurs from which the single heart tube develops (Allan and Kramer, 2010). The tube develops a series of constrictions which divides it into anatomical parts. From the cranial to the caudal ends these are, the truncus arteriosus, where the outflow tract of the heart develops, the bulbus cordis, between the truncus and the single ventricle, the ventricle, which is the largest part of the tube, the atrium, and the sinus venosus (Allan and Kramer, 2010). Further growth of the heart results in it ‘bending’ and ‘twisting’.

Towards the end of the fourth week post-fertilisation, endocardial cushions (intermediate bar) form on the dorsal and ventral walls of the atrioventricular canal. These masses of tissue are invaded by mesenchymal cells during the fifth week post-fertilisation. The atrioventricular

endocardial cushions approach each other and fuse, dividing the single atrioventricular canal into the right and left atrioventricular canals (Moore and Persaud, 2003). These canals partially separate the primordial atrium from the primordial ventricle, and the endocardial cushions function as atrioventricular valves (Moore and Persaud, 2003).

Septation of the single ventricle begins when a muscular ridge grows superiorly from the base of the ventricle towards the intermediate bar. This is termed the muscular part (pars muscularis) of the interventricular septum (Allan and Kramer, 2010). The growth of the pars muscularis stops short of the intermediate bar, leaving a gap between its superior end and the bar. This gap is the interventricular foramen.

The interventricular foramen will be closed by the formation of the membranous part (pars membranacea) of the interventricular septum (Allan and Kramer, 2010). The pars membranacea has a complicated development. It is formed by a downgrowth of the intermediate bar with contributions from the anterior and posterior truncal ridges (also known as bulbar ridges). Neural crest cells migrate into the truncal ridges and are therefore involved in the formation of the interventricular septum (Allan and Kramer, 2010). Because of its complicated developmental history, it is most often the membranous part of the interventricular septum which fails to develop normally. The pars muscularis and the pars membranacea together form the definitive interventricular septum. The interventricular septum divides the original single ventricular chamber into right and left ventricles.

Recent advances in the study of the development of the heart have drawn attention to a second cardiac plate in addition to the primary cardiogenic plate. This second cardiac plate which occurs slightly later in development is said to contribute to the formation of the right ventricular outflow tract and to the atrioventricular valves (endocardial cushion area) (Cai *et al*., 2003, Buckingham *et al*., 2005). In recent years, molecular studies on the development of the heart are now elucidating genes and transcription factors which are involved in heart development and maldevelopment. For example, the null mutation for the transcription factor GATA3 results in a severe defect in the outflow tract, such as a ventricular septal defect (Raid *et al*., 2009).

1.2 The anatomy of the normal interventricular septum

Anatomically, the interventricular septum is said to be divided into four regions viz. the inlet septum, the trabecular septum, the outlet septum (the latter two referred to as the muscular septum), and the membranous septum (Soto *et al*., 1980). However, the boundaries and the anatomical landmarks of the different parts of the interventricular septum are not clearly defined in the literature, as different authors use different landmarks to describe them. For this reason the widely used definitions used by Soto *et al*. (1980) have been quoted in this study.

The inlet septum is that portion of the septum that separates the tricuspid and mitral valves (Fig.1.1). The inlet septum merges with the trabecular septum and separates the finely trabeculated left ventricular apex from the more coarsely trabeculated right ventricular apical area. When viewed from the right ventricle, it extends from the annulus of the tricuspid valve to the chordal attachments of the tricuspid valve on to the papillary muscles (Soto *et al*., 1980).

The trabecular septum extends from the tricuspid valve attachments inferiorly to the apex and superiorly to the crista supraventricularis (supraventricular crest) (Fig.1.1). The trabecular septum also merges with the outlet septum which separates the right and left ventricular outflow tracts, being considerably more extensive on its right ventricular aspect than on the left (Soto *et al*., 1980). When viewed from the right ventricle, the trabecular septum extends from the inlet septum to the cardiac apex (Soto *et al*., 1980).

The outlet septum extends from the crista supraventricularis to the pulmonary valve (Fig.1.1) (Soto *et al*., 1980).

The membranous septum in the normal heart is a small structure, divided into two parts by the insertion of the septal leaflet of the tricuspid valve (an atrioventricular and an interventricular component). The membranous septum is situated inferior to the crista supraventricularis, posterior to the papillary muscle of the conus and immediately beneath the aortic valve. When viewed from the right ventricle it lies beneath the septal leaflet of the tricuspid valve and extends superiorly towards the aortic valve (Fig.1.1) (Soto *et al*., 1980).

In order to describe ventricular septal defects, it is necessary to understand the muscular trabeculae and the bands that are observed in the normal right ventricle. The normal pulmonary and tricuspid valves are separated by an extensive ledge termed the crista supraventricularis, which has two components (Soto *et al*., 1980). The component of the normal crista that separates the aortic and pulmonary valves is referred to as the infundibular septum. The second component, found between the pulmonary and tricuspid valves is termed, the

ventriculoinfundibular fold (parietal band). The extensive septal trabeculations of the right ventricle are referred to as the trabecula septomarginalis (Soto *et al*., 1980). The medial papillary muscle arises from the posterior limb of the trabecula septomarginalis.

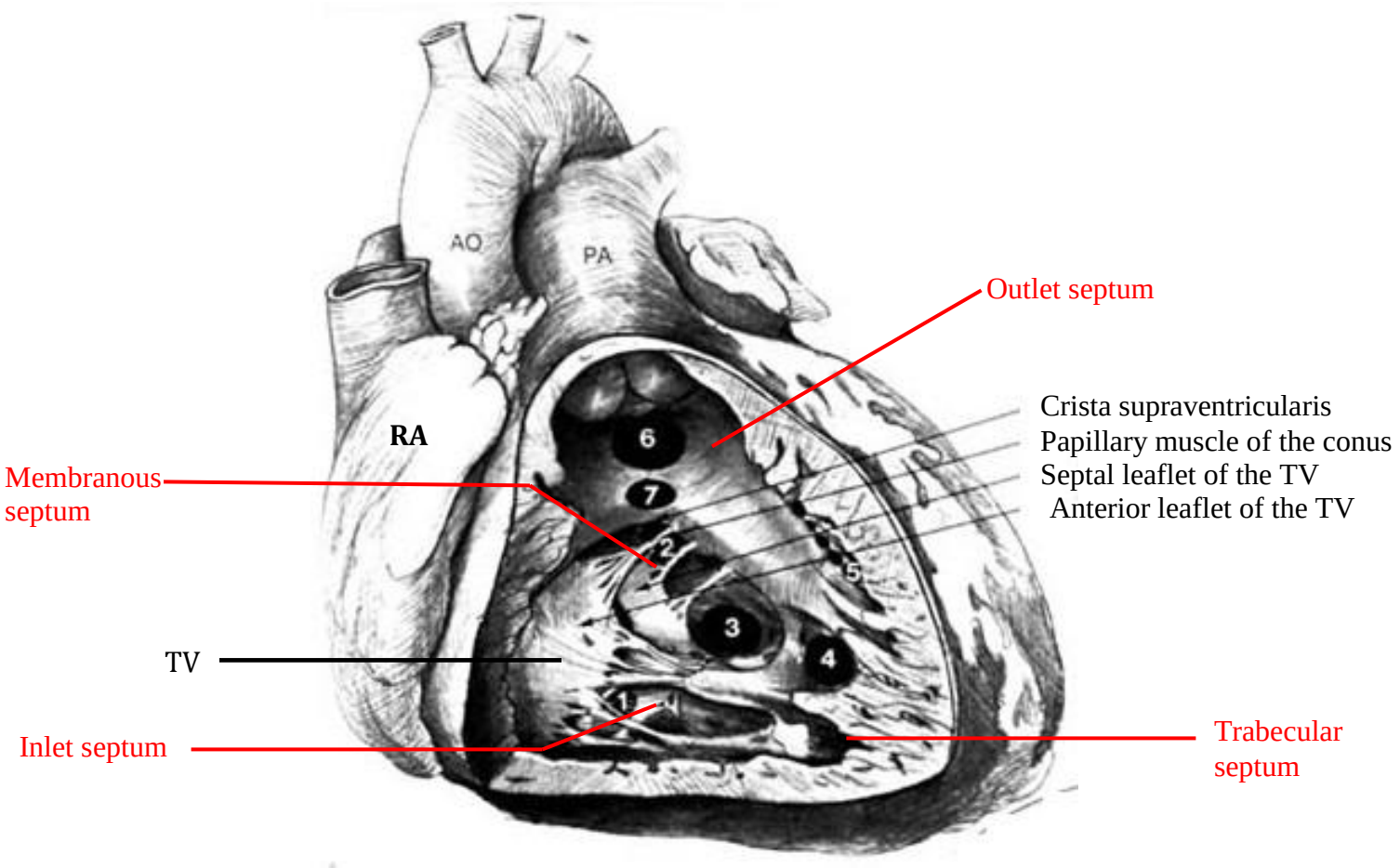


Figure 1.1: Classification of the interventricular septum and ventricular septal defects viewed from the right ventricle (Krabill, 2004)

1- inlet VSD

2- perimembranous VSD

3- central muscular VSD

4- apical muscular VSD

5- marginal muscular VSD

6- outlet VSD

7- midcristal VSD

AO- aorta

PA- pulmonary artery

RA- right atrium

TV- tricuspid valve

1.3 Ventricular septal defects

Major cardiac abnormalities generally develop during the fourth to the tenth week post-fertilisation (Trines and Hornberger, 2004). The incidence of ventricular septal defects as a single lesion varies from 1.5 to 3.5 per 1000 live births and from 4.5 to 7 per 1000 premature live births (Krabill, 2004). This defect is slightly more common in females than in males. It is the most common cardiac defect found in some chromosomal abnormalities (Trisomy 13-15, Trisomy 16-18, Trisomy 21). However, in 95% of cases it is not associated with a chromosomal defect. The etiology appears to be multifactorial with an interaction of hereditary predisposition and environmental factors (Krabill, 2004).

Although ventricular septal defects can occur as single or multiple lesions, the solitary ventricular septal defects account for approximately 20% of patients with congenital heart disease (Krabill, 2004), and are the most common acyanotic lesion. In recent years the frequency of spontaneous closure of ventricular septal defects has been well delineated. It is now clear that spontaneous closure of ventricular septal defects may occur in the first few years of life (Krabill, 2004). Ammash *et al.* (2001) in an American study, reported spontaneous closure of ventricular septal defects in 40% of children. Closure of the defect occurs as a result of muscular growth around the defect together with the growth of proliferative fibrous tissue or by a leaflet of the tricuspid valve adhering to the edge of the defect (Ammash *et al.,* 2001).

1.4 Classification of ventricular septal defects

Although ventricular septal defects may occur in any part of the interventricular septum, they occur more frequently in the perimembranous part. Incomplete closure of the interventricular foramen results from failure of the membranous part of the interventricular septum to develop. It occurs when the intermediate bar fails to fuse with the anterior and posterior truncal ridges and/or with the muscular part of the interventricular septum (Moore and Persuad, 2003).

While several classifications of ventricular septal defects have been proposed, most paediatric cardiologists prefer the classification of Soto *et al.* (1980) which is based on a paediatric study in Britain on 220 isolated or multiple ventricular septal defects excluding tetralogy of Fallot. A summary of this classification, more recently modified by the same authors is presented in Table 1.1 (Soto *et al*., 1980).

Table 1.1: Summary of the classification of ventricular septal defects (Soto *et al*., 1980)

Type of VSD	Synonym	% of VSD	Characteristics of defects
Membranous	Perimembranous Infracristal	80%	Situated adjacent to the membranous septum and can extend into the muscular tissue
Muscular Apical Central Marginal	"Swiss cheese" septum (When it occurs as multiple defects)	5-20%	Occurs in any combination of muscular locations
Inlet		8%	Not associated with defects of the AV valves and the common AV bundle does not pass beneath the defect
Outlet	Supracristal Conal Subpulmonary Subarterial	5-7%	Muscular support below the aortic valve may be deficient causing right coronary cusp prolapse into the VSD

VSD: ventricular septal defect; AV: atrioventricular

Perimembranous ventricular septal defects are the most common, occurring in 80% of surgical and autopsy cases (Soto *et al*., 1980). This defect may extend into the adjacent muscular tissue (Fig.1.1). Occasionally the septal leaflet of the tricuspid valve is deficient at its attachment to the membranous septum. This causes left ventricular to right atrial shunting (Soto *et al.,* 1980). A study conducted by Klass *et al.* (1990) in a South African population on 309 children, also found the perimembranous ventricular septal defect to be the most common type of ventricular septal defect (65%).

Muscular ventricular septal defects may occur as a single defect or maybe multiple and account for 5-20% of cases (Soto *et al.,* 1980). These muscular ventricular septal defects are further

divided based on their location (Soto *et al.,* 1980). Apical muscular ventricular septal defects are more common and are bordered by trabeculae and channels when viewed from the right ventricle (Fig.1.1). Central muscular ventricular septal defects are posterior to the trabecular septomarginalis in the mid-portion of the septum (Fig.1.1). Marginal muscular ventricular septal defects are located in the anterior septal free wall and are usually multiple with small channels (Soto *et al.,* 1980) (Fig.1.1). Muscular ventricular septal defects were found in 4% of cases in the South African study by Klass *et al.* (1990). Multiple muscular ventricular septal defects can occur in any combination of these muscular locations and may even include defects of the membranous septum. "Swiss cheese" septum is the term frequently used to describe these defects (Soto *et al.,* 1980).

Inlet ventricular septal defects constitute 8% of ventricular septal defects. They differ from true atrioventricular septal defects (endocardial cushion defects) in that they are not associated with defects of the atrioventricular valves and the common atrioventricular bundle does not pass beneath the defect (Soto *et al.,* 1980).

Ventricular septal defects in the outlet septum constitute 5-7% of ventricular septal defects. In Japan the incidence of these defects can be as high as 30% (Tatsuno *et al.,* 1973a). No outlet ventricular septal defects were reported in a South African study by Klass *et al*. (1990). The ethnic similarity (between Blacks and Whites) of the anatomical type of ventricular septal defect in the South African paediatric population contrasts with the situation in Japan where there is a high prevalence of outlet ventricular septal defects. In outlet ventricular septal defects, the muscular support below the aortic valve may be deficient causing the right aortic coronary cusp

to prolapse into the ventricular septal defect (Tatsuno *et al*., 1973b). This results in aortic insufficiency and a mild to moderate degree of right ventricular outflow tract obstruction.

Recent advances in ultrasound technology have led to prenatal diagnosis of cardiac disease (Trines and Hornberger, 2004), and currently echocardiography is being used more frequently in clinical diagnosis.

1.5 Prolapse of the aortic cusp and aortic regurgitation

The association of aortic regurgitation with ventricular septal defects is well known. When the aortic regurgitation appears in infancy, in the majority of cases it is due to prolapse of the aortic cusp (Somerville *et al.,* 1970). Somerville *et al*. (1970) propose that another more likely explanation for aortic regurgitation which occurs with a ventricular septal defect is a congenital abnormality of the aortic valve which becomes susceptible to bacterial infection. As a result, the function of the aortic valve deteriorates resulting in aortic regurgitation.

Some authors (Nadas *et al.,* 1964; Van Praagh and McNamara, 1968) believe that prolapse of the right coronary cusp and consequently the aortic regurgitation is related to the site of the ventricular septal defect. A difference in the site of the ventricular septal defect associated with prolapse of the right coronary cusp has been reported in the literature. In Caucasians (British and American reports) the ventricular septal defect is most commonly the perimembranous type (Nadas *et al.*, 1964; Van Praagh and McNamara, 1968; Somerville *et al*., 1970). In contrast, Japanese patients have the ventricular septal defect situated in the subarterial (supracristal)

position (Tatsuno *et al.*, 1973a). The aortic leaflet which most commonly prolapses into the ventricular septal defect thus causing aortic regurgitation is the right coronary cusp of the aortic valve, the non-coronary cusp to a lesser degree and rarely the left coronary cusp (Nadas *et al.*, 1964). In the British study by Somerville *et al.* (1970), 20 patients presented with aortic regurgitation with ventricular septal defects. Prolapse of the right coronary cusp was documented as the primary cause of aortic regurgitation in only four of the 20 cases (Somerville *et al.*, 1970).

In cases of ventricular septal defects, aortic regurgitation develops at some time between the second to tenth years of life (Nadas *et al.*, 1964). In the study by Nadas *et al.* (1964) the most common time of appearance of aortic regurgitation was between three and eight years of age. Based on their findings, the authors concluded that ventricular septal defects with aortic regurgitation are not a congenital defect, as aortic regurgitation does not present at birth or during the first year of life. During this early period only the ventricular septal defect of small to moderate size is detected by the presence of a loud systolic murmur (Nadas *et al.*, 1964).

Aortic regurgitation increases in severity with an increase in age. It follows thus, that early closure of the defect may prevent or cure aortic regurgitation (Somerville *et al.*, 1970). Surgery for closure of the ventricular septal defect as well as for repair of the aortic valve is necessary in order for the patient to improve. Closure of the ventricular septal defect alone was not successful in curing the aortic regurgitation in the British study conducted by Somerville *et al.* in 1970. However, it was noted that the group of patients presented in the latter study were older and had long standing aortic regurgitation.

Tatsuno *et al.* (1973b) assessed the etiological determinants of a prolapsed aortic valve into a ventricular septal defect. Two intrinsic factors appear to be involved. The first is a lack of anatomical support for the valve and the second deals with hemodynamics that may augment prolapse (Tatsuno *et al*., 1973b). In the case of supracristal ventricular septal defects, prolapse of the aortic valve into the ventricular septal defect is due to a deficiency in the anatomical structures that support the aortic valve annulus and the sinus of Valsalva (Tatsuno *et al*., 1973b). Normally the sinus of Valsalva and the annulus of the aortic valve are supported by the parietal band on the right ventricular side. In some of the cases with ventricular septal defects this parietal band is deficient, resulting in prolapse of the right coronary cusp of the aortic valve and the sinus of Valsalva into the ventricular septal defect with subsequent aortic regurgitation (Tatsuno *et al*., 1973b). In patients with infracristal ventricular septal defects, two anatomical anomalies contribute to the prolapse of the aortic valve into the ventricular septal defect (Tatsuno *et al*., 1973b). One is the deficient parietal band as for the supracristal defects, and the other is due to abnormal development of the sinus of Valsalva.

Why do some patients with ventricular septal defects develop aortic regurgitation whereas most do not? Nadas *et al.* (1964) concluded that aortic regurgitation is an acquired complication of the ventricular septal defect and is not a congenital abnormality. Van Praagh and McNamara (1968) concluded from their study of 11 post-mortem cases, that aortic regurgitation is directly related to under-development of the aortic commissure in the cases of the infracristal defects, while in the cases of supracristal defects, the aortic regurgitation is due to the conal septal portion of the crista supraventricularis being absent or deficient. Their view concurs with that of Tatsuno *et al.* (1973).

The surgical management of the supracristal and infracristal types of ventricular septal defects differs; therefore their differential diagnosis is of clinical importance (Van Praagh and McNamara, 1968).

1.6 Left ventricular non-compaction associated with ventricular septal defects

Ventricular non-compaction is a rare congenital cardiomyopathy characterized by prominent trabeculations and deep intratrabecular recesses (Freedom *et al*., 2005). Non-compaction of the left ventricle is seen most frequently in the structurally normal heart. However, increasing recognition of this lesion shows that it can occur in association with different forms of congenital heart disease (Freedom *et al*., 2005). Morphologically, the papillary muscles are not well developed in these patients, and the non-compacted internal myocardial layers consist of more than 50% of the ventricular wall thickness (Ozgur *et al.*, 2011). Ventricular non-compaction constitutes approximately 9% of all childhood cardiomyopathies (Freedom *et al*., 2005). If ventricular non-compaction does not co-exist with other cardiovascular pathologies it is called ‘isolated’ ventricular non-compaction. Although the left ventricle is most commonly affected (62%), both ventricles can be affected in some cases (22–38%) (Ozgur *et al.*, 2011). When the left ventricle is involved, the sites most commonly affected are the apical, inferior and lateral parts respectively. Increased awareness and the use of modern ultrasound technology have resulted in increased detection of the morphological features of left ventricular non-compaction in routine clinical practice (Ichida, 2009). The echocardiographic criteria for left ventricular non-compaction is a ratio of greater than 2:1 between the non-compacted (internal) and compacted (external) areas of the left ventricle.

The clinical manifestation of left ventricular non-compaction is highly variable, ranging from no symptoms to a progressive deterioration in cardiac function that results in congestive heart failure, arrhythmias, thromboembolic events and sudden cardiac death (Ichida, 2009). Up to 40% of left ventricular non-compaction patients have evidence of familial disease, based on clinical assessment of their family members (Ichida, 2009). Left ventricular non-compaction has been linked to mutations in several genes, including LIM domain binding protein 3(*ZASP*), α-dystrobrevin (*DTNA*), tafazzin (*TAZ/G4.5*), lamin A/C (*LMNA*), and genes encoding the sarcomeric proteins, β-myosin heavy chain (*MYH7*), α-cardiac actin (*ACTC*), and cardiac troponin T (*TNNT2*) (Ichida, 2009).

Co-existence of left ventricular non-compaction and "Swiss cheese-like" multiple ventricular septal defects may especially explain the mal-development of the ventricular myocardium as the pathogenesis (Freedom *et al*., 2005). In 2009, Ichida investigated 103 Japanese patients with left ventricular non-compaction. In his study only two children were identified with "Swiss cheese-like" multiple ventricular septal defects.

1.7 The anatomical hallmarks of tetralogy of Fallot

Ventricular septal defects are also part of tetralogy of Fallot. Although the same classification for the ventricular septal defect is used for tetralogy of Fallot there are major fundamental differences. It is more than one hundred years since Etienne-Louis Arthur Fallot described tetralogy of Fallot in the journal Marseille Medicale, the clinical-pathological correlates of "la maladie bleue" (Fallot, 1888).

The tetralogy of Fallot can be diagnosed simply by noting the association of its four individual features namely, a large ventricular septal defect, deviation of the aorta to the right (overriding aorta), concentric hypertrophy of the right ventricle and right ventricular outflow tract obstruction (Fallot, 1888). However it is attractive to search for some readily recognizable morphological keystone that unites the overall entity (Goor and Lillehei, 1995). Anderson and Tynan (1988) and later Goor and Lillehei (1995) believed that such a landmark is provided by the abnormal antero-cephalad location of the outlet septum, such that it is malaligned relative to the rest of the ventricular septum and is attached exclusively within the morphologically right ventricle. This feature is readily recognized on cross-sectional echograms, angiograms or magnetic resonance images and by surgical observation. It is generally accepted that the hypertrophy of the right ventricle is the haemodynamic consequence of the anatomical derangement, and that the degree of hypertrophy becomes greater with increasing age. The anatomical focus, therefore, can be directed to the malaligned outlet septum (Anderson and Tynan, 1988).

The tetralogy of Fallot accounts for 4-10% of all congenital heart diseases and is the most common cyanotic congenital heart disease, both in children and in adults (Hoffman and Kaplan, 2002). Males and females are equally affected. The risk of recurrence in siblings is about 3%. A deletion of chromosome 22q11 is found in 15% of patients who have tetralogy of Fallot (Hoffman and Kaplan, 2002). Typically, there is only a heart murmur in early infancy, with cyanosis becoming obvious in early childhood due to right to left shunting across the ventricular septal defect because of the right ventricular outflow tract obstruction (Hoffman and Kaplan,

2002). Intra-cardiac repair is feasible, with reasonably good long-term results (Hoffman and Kaplan, 2002).

There is wide variation with regard to the degree and site of right ventricular outflow obstruction, pulmonary artery size and the degree of aortic overriding. The ventricular septal defect usually occurs in the perimembranous region, but is located in the infundibular septum in 20% of Asian patients with tetralogy of Fallot (Hoffman and Kaplan, 2002). It may be accompanied by additional anomalies such as a patent ductus arteriosus, pulmonary atresia, hypoplasia or stenosis of the pulmonary artery, major aortopulmonary collateral arteries, absent pulmonary valve, anomalies of the coronary artery and anomalies of the aortic arch i.e. right sided instead of left sided in 25% of cases (Hoffman and Kaplan, 2002).

1.8 Double chambered right ventricle in association with ventricular septal defects

Double chambered right ventricle is an uncommon congenital heart disease, characterized by division of the right ventricle by hypertrophied muscle bands into two parts, the proximal high pressure chamber and the distal chamber with low pressure (Telagh *et al.*, 2008). In addition, it is frequently associated with ventricular septal defects. The initial clinical presentation of a double chambered right ventricle varies between a cardiac murmur, cyanosis or congestive heart failure. This variable presentation thus depends on the presence, site and size of the associated ventricular septal defect together with the severity of the obstruction within the right ventricle (Telagh *et al.*, 2008). In most cases of double chambered right ventricle, the aberrant muscle band causes obstruction of pulmonary blood flow with pressure overload in the right ventricle

(Telagh *et al.*, 2008). Thus these patients with cyanosis are clinically very similar to those with tetralogy of Fallot.

The developmental mechanism of double chambered right ventricle is unclear. Rowland *et al.* (1975) proposed that improper expansion of the bulboventricular junction may result in incomplete fusion of the bulbar and endocardial cushion elements that normally close the superior portion of the ventricular septum. This results in right ventricular subdivision. According to their theory, all components of the right ventricle superior to the moderator band, as well as a distal segment of the left ventricular outflow tract are of bulbar origin. The authors further argue that the right ventricular obstruction in patients with double chambered right ventricle is not due to the moderator band (as this normally arises from the inferior portion of the septal band), but rather results from an elevated origin of a hypertrophied moderator band from the septal band (which originates near the crista supraventricularis) (Rowland *et al.*, 1975).

The site of right ventricular obstruction as seen in tetralogy of Fallot typically occurs at the mouth of the subpulmonary infundibulum while that seen in double chambered right ventricle, is located within the apical trabecular compartment (Telagh *et al.*, 2008). Due to the developmental and progressive nature of double chambered right ventricle, early surgical intervention to correct the obstruction and to close the ventricular septal defect is warranted (Telagh *et al.*, 2008).

1.9 Overall aims of the study

Although the literature is replete with numerous studies describing ventricular septal defects, very little is documented about them in the different population groups in South Africa. A wider knowledge of the spectrum of these ventricular septal defects may be useful to clinicians in deciding on the correct surgical approach.

This study therefore aimed to:

- Describe the size, boundaries and relations of the interventricular septum in neonatal cadaveric hearts
- Document the ventricular septal defects seen in post-mortem specimens of hearts
- Document the types of isolated ventricular septal defects seen at surgery in Johannesburg provincial hospitals during the period 2001 to 2004 and to compare
- The types of ventricular septal defects, and their incidence which occur in tetralogy of Fallot and double chambered right ventricle.

2. MATERIALS AND METHODS

This study is divided into two parts, namely an anatomical study and a retrospective clinical study.

2.1 Anatomical study

In order to document the size, boundaries and the relations of the different parts of the interventricular septum, the hearts of 11 neonates were dissected. All the neonates had been fixed in a formalin/alcohol solution in the School of Anatomical Sciences, Faculty of Health Sciences, University of the Witwatersrand. The neonates form part of a collection of human cadaveric specimens in the School of Anatomical Sciences and have been dissected in accordance with the Human Tissues Act of 1985 of the Republic of South Africa. A Human Ethics Committee waiver was also received from the University of the Witwatersrand.

The sex and the population group of all the cadaveric specimens were documented. The sex organs of all the neonates were fully developed and were visible externally. The age of the neonates was assessed by comparing crown-rump length, foot length and bi-parietal breadth from an age estimation chart (Moore and Persaud, 1998).

Crown-rump length was taken from the crown of the head to the rump of the neonate, using a steel measuring tape. Foot length was taken from the second toe to the heel on the left foot using a sliding caliper. Bi-parietal breadth was taken from euryon to euryon using a spreading caliper. The maximum breadth of the skull was determined by moving both ends of the spreading caliper

back and forth on the sides of the skull until the maximum breadth was located. All measurements were recorded in millimeters to one decimal place. Repeat measurements were taken and averages were used. These averages were then compared to values on an age estimation chart (Moore and Persaud, 1998) and an approximate age of each neonate was determined.

The dissection method described by Kieser and Allan (2001) was followed when removing the heart from the thoracic cavity. Micro-dissection of the neonatal hearts was performed under a stereomicroscope. The interventricular septum was exposed through the right ventricle and photographed with a digital camera. Prior to dissection, and at every stage of the dissection, all structures were identified and recorded and digital photographs were taken.

A sliding caliper was used to take measurements of the components of the neonatal interventricular septum. Measurements were taken in millimeters to one decimal place. Repeat measurements were taken on different days and at different times on all specimens to ensure accuracy. The raw data appears in tabulated form in appendix A.

Boundaries of the components of the interventricular septum viewed from the opened right ventricle

In order to facilitate accuracy when measuring the size of the different components of the interventricular septum, the following landmarks (Soto *et al*., 1980) were used to demarcate the different components of the septum in each of the neonatal hearts:

1) Inlet septum – from the annulus of the tricuspid valve to the chordal attachments of the tricuspid valve on to the papillary muscles (Fig.2.1).
2) Outlet septum - from the crista supraventricularis to the pulmonary valve (Fig.2.1).
3) Muscular septum - inferior to the crista supraventricularis, posterior to the papillary muscle and inferior to the aortic valve (Fig.2.1).
4) Perimembranous septum – inferior to the crista supraventricularis, near the pulmonary valve, near the base of the septal leaflet of the tricuspid valve on the right and the mitral valve on the left (Fig.2.1).

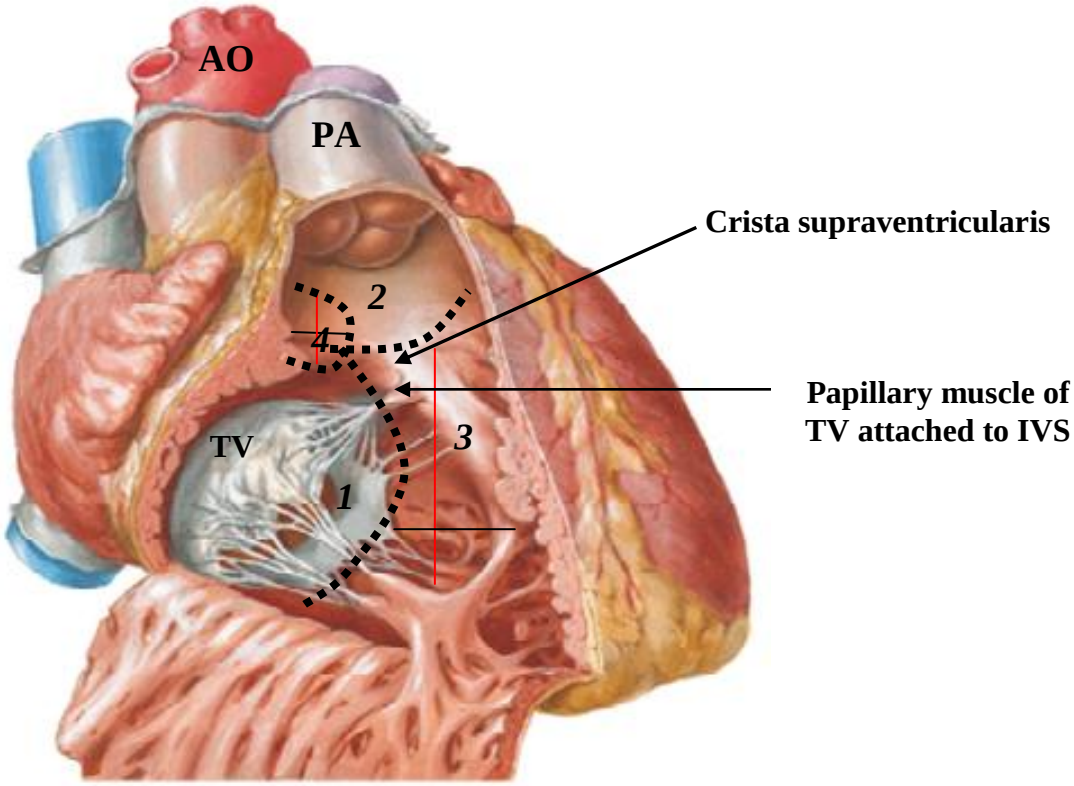


Figure 2.1: Boundaries of the components of the interventricular septum (IVS)
Demarcations (-----) indicate: 1) inlet septum; 2) outlet septum; 3) muscular septum; 4) perimembranous septum. Ao= aorta; PA= pulmonary artery; TV= tricuspid valve
Red lines (____) indicate length and Black lines (____) indicate breadth
(Adapted from Netter, 1997)

Once the landmarks were clearly defined and documented, the following parameters were used to measure the size of each of the different components of the interventricular septum on each of the dissected neonatal hearts:

1) Maximum length of the inlet septum
2) Maximum length of the outlet septum
3) Maximum length of the muscular septum
4) Maximum length of the perimembranous septum
5) Maximum breadth of the muscular septum
6) Maximum breadth of the perimembranous septum

In addition, seven heart specimens from Professor Solly Levin's post mortem collection housed in the Division of Paediatric Cardiology, Faculty of Health Sciences, University of the Witwatersrand were studied in order to gain an understanding of ventricular septal defects. Four of these specimens were diagnosed with an isolated ventricular septal defect, and three displayed the features of tetralogy of Fallot.

2.2 Retrospective study of clinical records

Fifty cases of isolated ventricular septal defects, forty two cases of ventricular septal defects with tetralogy of Fallot and eight cases of ventricular septal defects with double chambered right ventricle who had undergone open heart surgery at the Charlotte Maxeke Hospital (Johannesburg general) were used for the study. The operation reports together with the clinical case reports

provided by Paediatric Cardiologists to the Cardiothoracic Surgeons from the three main referral hospitals in Johannesburg (Charlotte Maxeke, Raheema Moosa mother and child {Coronation} and Chris Hani Baragwanath) were used to collect this data retrospectively from 2001 to 2004. Ethical clearance to undertake this study was obtained from the Human Ethics Committee of the University of the Witwatersrand (Ethics clearance number-R14/49). The collection of data from the patient's files included clinical information, echocardiographic and catheterization reports, as well as surgical operative notes. All the reports and notes were reviewed twice on different days to ensure that the data had been recorded accurately.

Prenatal history

The data were categorized and documented as follows:

- Normal birth
- Complicated birth (e.g. premature birth, birth by caesarian section and breech delivery).

Demographics

The following information was recorded:

a) Population affinity- e.g. Black, White, Coloured and Indian

b) Sex

c) Age in years and months. The infants and children were grouped into age cohorts in order to afford analysis of the data. The patients with isolated ventricular septal defects, ventricular septal defects occurring in tetralogy of Fallot and the ventricular septal defects occurring in double

chambered right ventricle were subdivided into the following three age groups and will be referred to in these groups in the text:

- Group A: less than 2 years old
- Group B: 2-4 years
- Group C: greater than 4 years

d) Weight in kilograms (from the catheterization reports) and

e) Height in meters (from the catheterization reports)

Additional information was documented as follows:

First presentation of the ventricular septal defect and the subsequent operation details

First presentation of the ventricular septal defect:

- Reason for initial ventricular septal defect diagnosis
- Reason for delaying repair

Cardiac operations prior to ventricular septal defect repair

All previous cardiac operations, which any of the patients had undergone, prior to the operation for the ventricular septal defect were documented.

Other features associated with the ventricular septal defect

a) Type and location of the ventricular septal defect i.e. Perimembranous, muscular, outlet, inlet
b) Size of the ventricular septal defect in millimeters. This was recorded from the echocardiogram (as documented in the clinical case reports).
c) Haemodynamic consequence of the ventricular septal defect- the following data was recorded:
    - pulmonary to systemic flow ratio (Qp:Qs)
    - systolic pulmonary artery pressure
d) Additional cardiac abnormalities

All the raw data in the study is listed in Appendix A.

A statistician was consulted for the statistical component of the study. Due to the small sample size (e.g. 50 in one group, 42 in another and a further subgroup of eight) little statistical analysis could be conducted on the data besides descriptive statistics. The research thus presents a series of descriptive analyses.

3. RESULTS

3.1 Anatomical study

3.1.1 Cadaveric study from the School of Anatomical Sciences

The age assessment of the neonates was carried out by comparing crown-rump length, foot length and bi-parietal breadth from an age estimation chart. These calculated ages of the neonates which were studied are tabulated in Table 3.1.

Seven of the 11 normal neonates were female and four were males. All the neonates were of the Black population. The calculated age of the neonates ranged from 24 to 40 weeks (Table 3.1). Three of the 11 neonatal hearts were excluded from the sample, as the anatomical landmarks of the different components of the interventricular septum could not be accurately defined. The results of the morphometric analysis of the components of the interventricular septum are shown in Table 3.2.

In the remaining eight neonatal hearts, the interventricular septum was divided into four regions viz. perimembranous, muscular or trabecular, inlet and outlet (Fig.3.1).

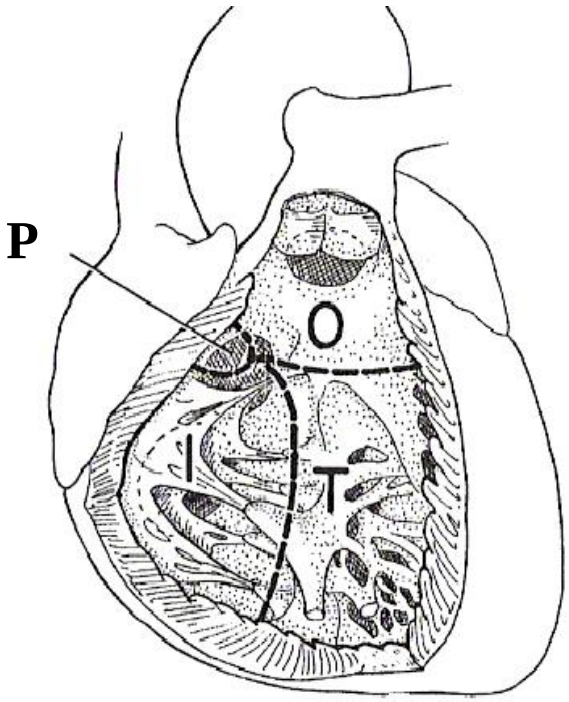


Figure 3.1: Divisions of the interventricular septum
Adapted from a presentation by V. Hunter, Division of Paediatric Cardiology
(P: perimembranous, T: muscular or trabecular, I: inlet, O: outlet)

The perimembranous septum in all the normal neonates was situated inferior to the crista supraventricularis, posterior to the papillary muscle of the conus and immediately beneath the aortic valve. When viewed from the right ventricle, the perimembranous septum lay beneath the septal leaflet of the tricuspid valve and extended up to the aortic valve. The perimembranous septum and the muscular septum made up the largest component of the interventricular septum with the mean length of the perimembranous septum ranging from 4.0 mm to 12.1 mm and the breadth from 16.6 mm to 23.5 mm (Table 3.2). The mean length and breadth of the perimembranous septum was found to be greater in the neonate aged between 38 to 40 weeks

(12.1 mm and 23.3 mm respectively) when compared to that of the neonate aged between 24- 26 weeks (5.7 mm and 19.5 mm respectively) (Table 3.2).

The muscular septum extended from the tricuspid valve attachment towards the apex and superiorly towards the crista supraventricularis. It extended from the inlet septum to the cardiac apex when viewed from the right ventricle. The muscular septum had a mean length ranging from 16.0 mm to 18.2 mm and a mean breadth ranging from 15.9 mm to 19.0 mm (Table 3.2). There was no difference between the mean breadth of the muscular septum at 38 to 40 weeks and at 24 to 26 weeks of age (Table 3.2).

The inlet septum separated the tricuspid and mitral valves. When viewed from the right ventricle, the inlet septum extended from the annulus of the tricuspid valve to the chordal attachments of the tricuspid valve and on to the papillary muscles.

The outlet septum extended from the crista supraventricularis to the pulmonary valve.

The inlet septum and outlet septum contributed the smaller components of the remainder of the interventricular septum with mean lengths which ranged from 3.6 mm to 9.6 mm and from 3.3 mm to 7.9 mm respectively (Table 3.2). The length of the outlet and inlet septum was greater at 38 to 40 weeks than at 24 to 26 weeks of age (Table 3.2). The breadth of the inlet and outlet septum was not calculated, as the anatomical landmarks could not be identified to facilitate accurate measurements on the neonatal cadavers.

3.1.2 Post-mortem study of heart specimens from the Division of Paediatric Cardiology at the Charlotte Maxeke Academic Hospital

In the four random postmortem specimens which had perimembranous ventricular septal defects, these defects were located in the membranous part of the septum and were beneath the crista supraventricularis and posterior to the papillary muscle of the conus (as viewed from the right ventricle) (Fig. 3.2).

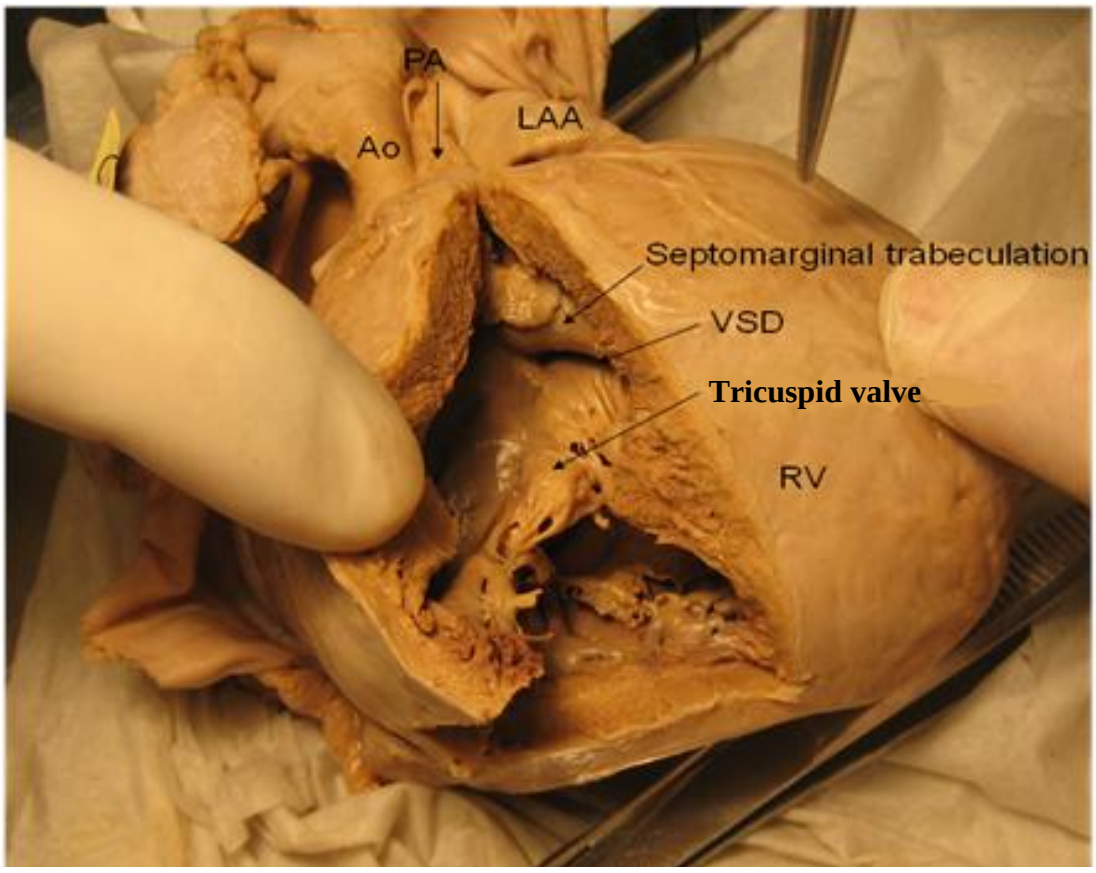


Figure 3.2: Postmortem specimen demonstrating a perimembranous ventricular septal defect viewed from the right ventricle
(Adapted from V. Hunter's presentation -Senior Paediatric Cardiac Technologist, Division of Paediatric Cardiology at the Charlotte Maxeke Hospital)
(Ao: aorta, PA: pulmonary artery, LAA: left atrial appendage, VSD: ventricular septal defect, RV: right ventricle)

In one of the four cases (case one), that of a 16 day old male diagnosed with Down's syndrome the left anterior descending coronary artery was seen demarcating the left ventricle from the right ventricle (Fig.3.3). The right ventricle was larger than the left and was very trabeculated (more than usual) (Fig.3.3). The left atrial appendage was smaller than usual. A patent foramen ovale and patent ductus arteriosus were observed. The perimembranous ventricular septal defect measured five millimeters in diameter (Fig.3.4).

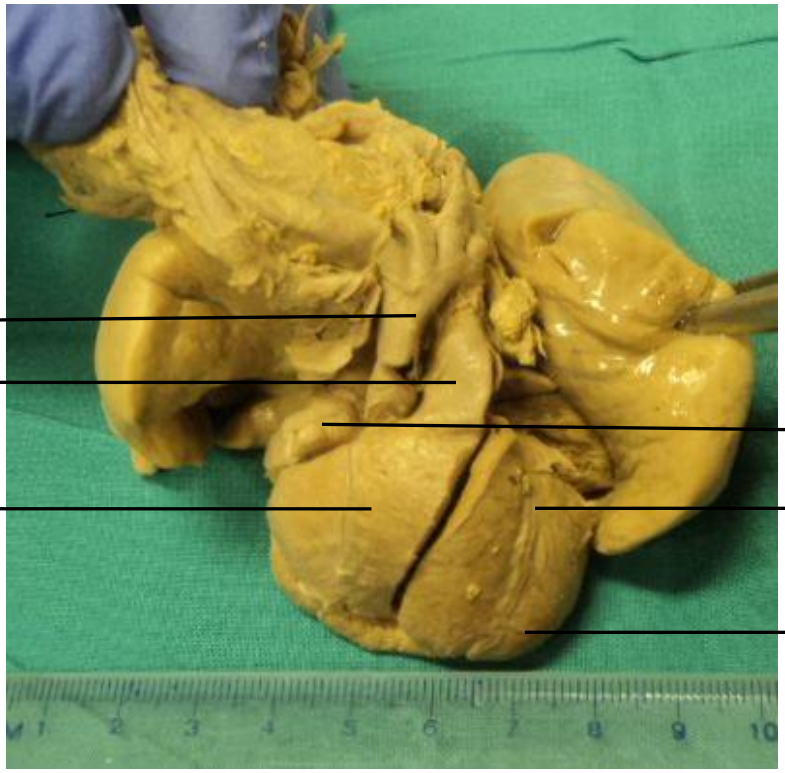


Figure 3.3: Case 1- Frontal view of a heart demonstrating the right and left ventricles and the major arteries
(Photographed from Professor Levin's post mortem collection)
Note: Enlarged main pulmonary artery trunk

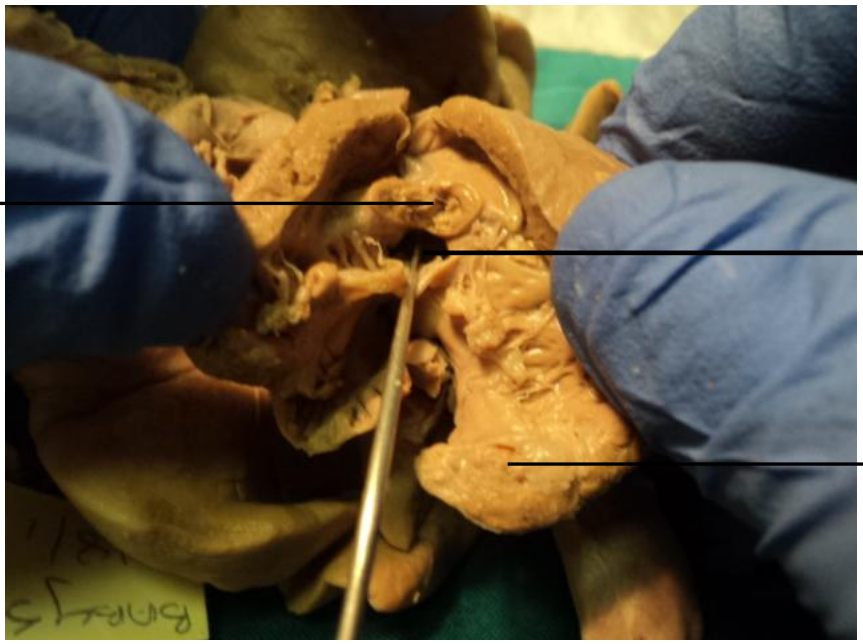


Figure 3.4: Case 1- Right ventricular view of the perimembranous ventricular septal defect (probe inserted)
(Photographed from Professor Levin's post mortem collection)
Note: Damaged crista supraventricularis

In case two, no clinical information was available on record. The post mortem specimen showed a grossly enlarged heart suggesting an associated dilated cardiomyopathy (Fig.3.5). Both ventricles were dilated with some associated hypertrophy. A perimembranous ventricular septal defect (11mm) was present and a further diagnosis of non-compaction of the left ventricle was made due to prominent trabeculations in that ventricle (Fig.3.6).

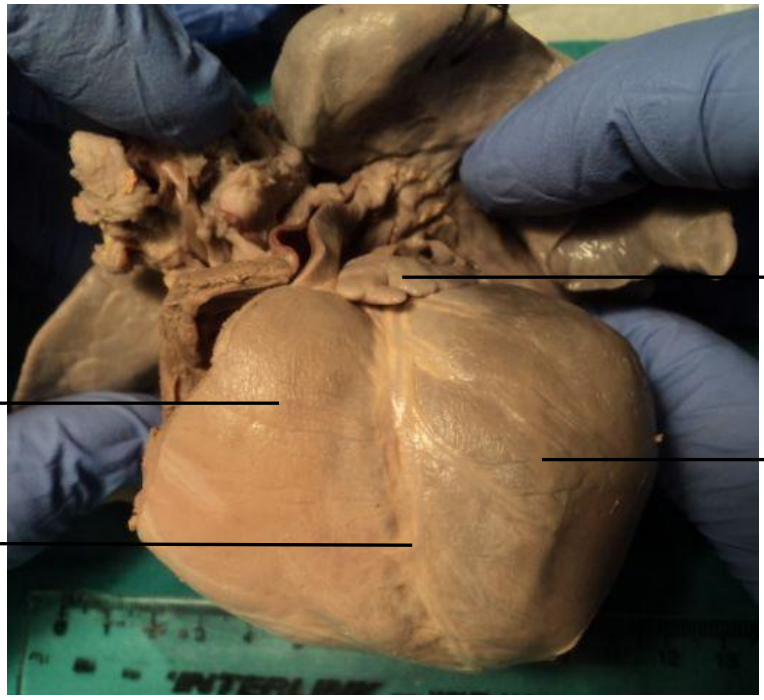


Figure 3.5: Case 2- Frontal view of the dilated heart showing dilated left and right ventricles (dilated cardiomyopathy)
(Photographed from Professor Levin's post mortem collection)

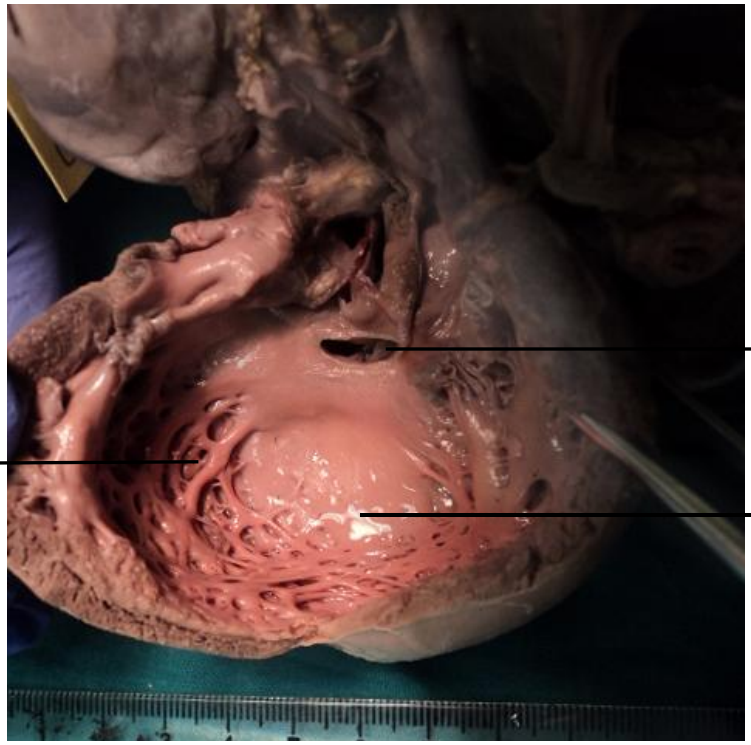


Figure 3.6: Case 2-Photographic representation of a perimembranous ventricular septal defect and non-compaction (prominent trabeculations) [Photographed from Professor Levin's post mortem collection]
Note: Dilated left ventricle

In case three, the tricuspid valve was observed with nodules on its edges (Fig.3.7). The left atrium was grossly dilated. Tight mitral valve stenosis was observed with shortened chordae. The perimembranous ventricular septal defect measured eight millimeters on the left ventricular side (Fig.3.8).

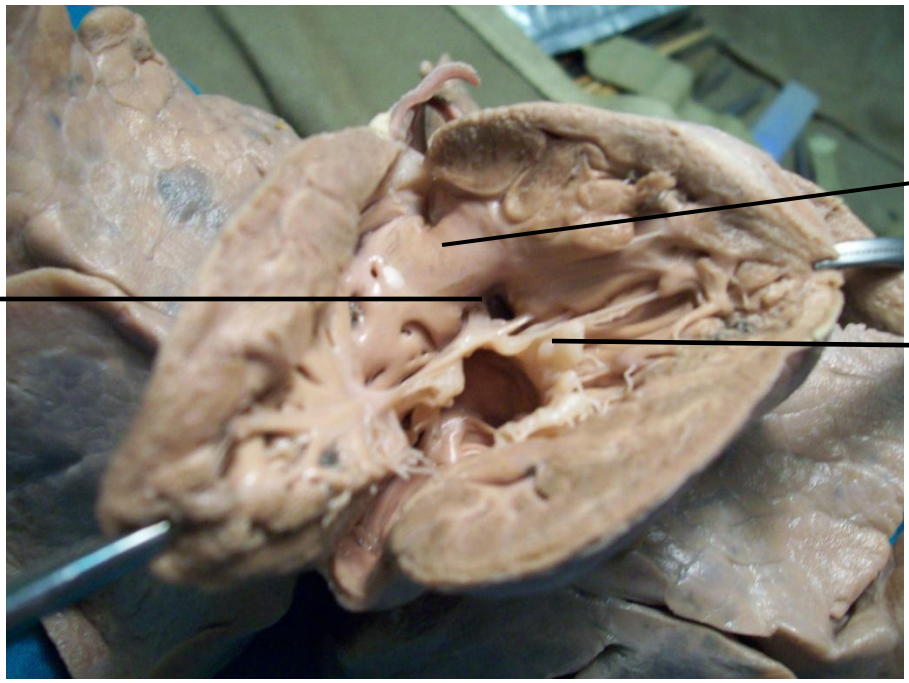


Figure 3.7: Case 3- Perimembranous ventricular septal defect viewed from the right ventricle
[Photographed from Professor Levin's post mortem collection]
Note: Nodule on edge of the tricuspid valve

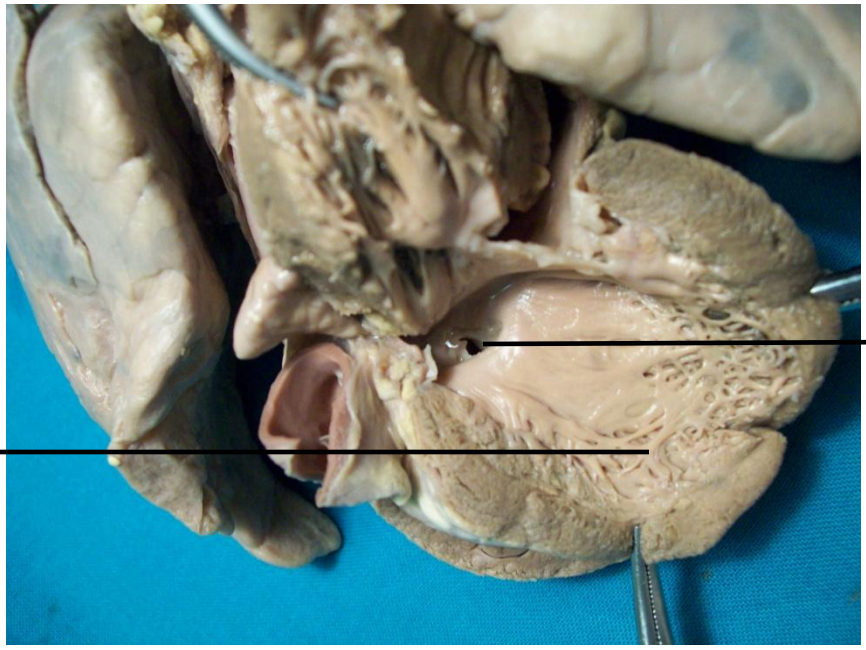


Figure 3.8: Case 3- Perimembranous ventricular septal defect viewed from the left ventricle
[Photographed from Professor Levin's post mortem collection]

In case four, the perimembranous ventricular septal defect measured six millimeters on the left ventricular side (Fig 3.9). Blood cysts were observed on the tricuspid valve (Fig. 3.10). The left atrium was grossly dilated.

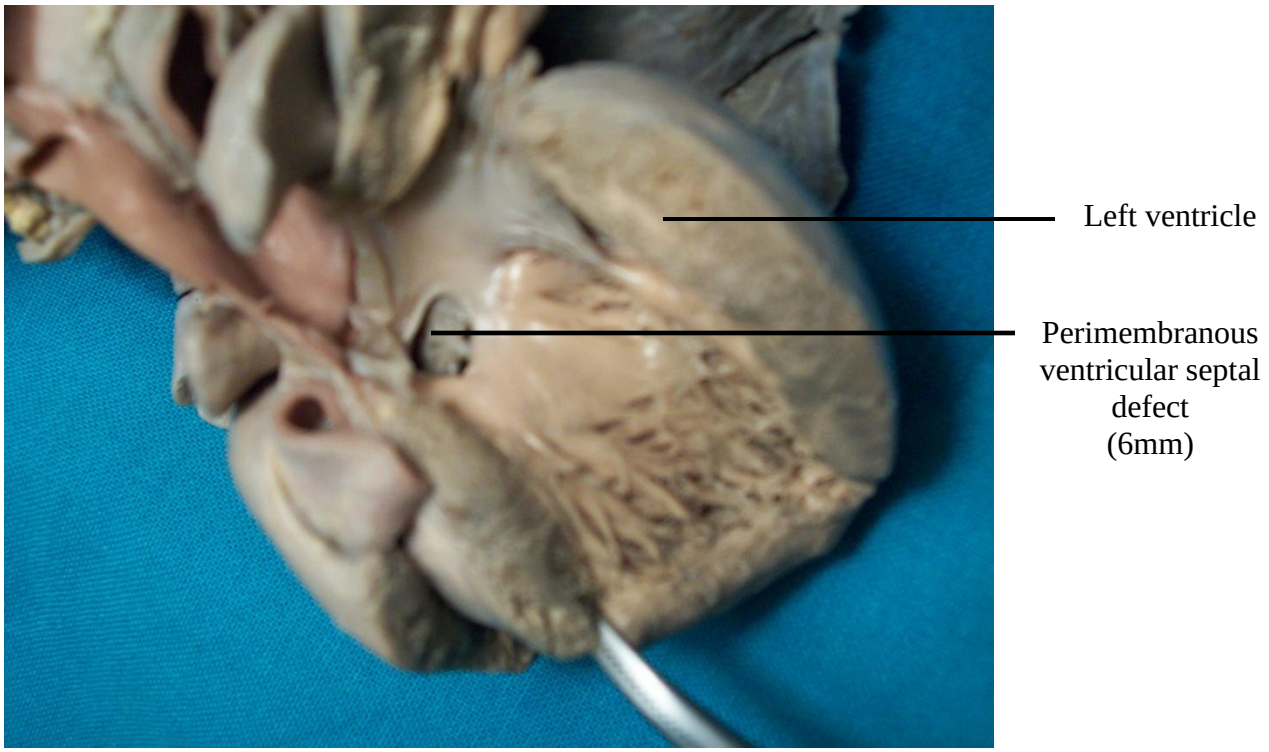


Figure 3.9: Case 4- Perimembranous ventricular septal defect viewed from the left ventricle
[Photographed from Professor Levin's post mortem collection]



Figure 3.10: Case 4- Perimembranous ventricular septal defect viewed from the right ventricle with blood cyst on the tricuspid valve [Photographed from Professor Levin's post mortem collection]

In the three postmortem specimens with tetralogy of Fallot, the pulmonary artery trunk was noticeably smaller than the aorta. The right ventricles were hypertrophied and enlarged, and the ventricular septal defects were in the membranous area in all three cases.

Case five was a neonate diagnosed with tetralogy of Fallot born by caesarian section at 33 weeks. The examination of the post-mortem specimen revealed a small heart, a small main pulmonary artery trunk (four millimeters), a large overriding aorta (nine millimeters) (Fig. 3.11), a large perimembranous ventricular septal defect (six millimeters) (Fig. 3.12) and narrowing of the right ventricular outflow tract. The right ventricle was severely hypertrophied. Other abnormalities included a left diaphragmatic hernia, hypoplastic left lung, cleft palate, undescended testis and a bifid left thumb.

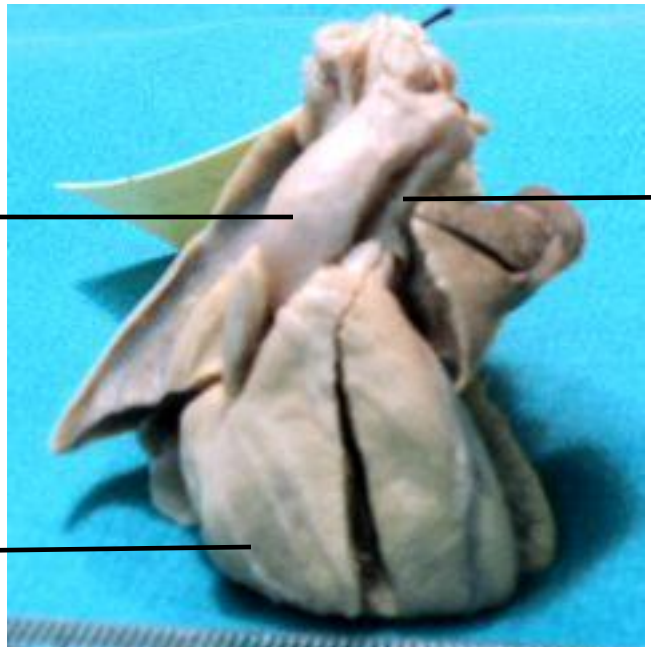


Figure 3.11: Case 5- Frontal view of a post-mortem specimen of a case diagnosed with tetralogy of Fallot displaying a large overriding aorta and a small main pulmonary artery trunk [Photographed from Professor Levin's post mortem collection]

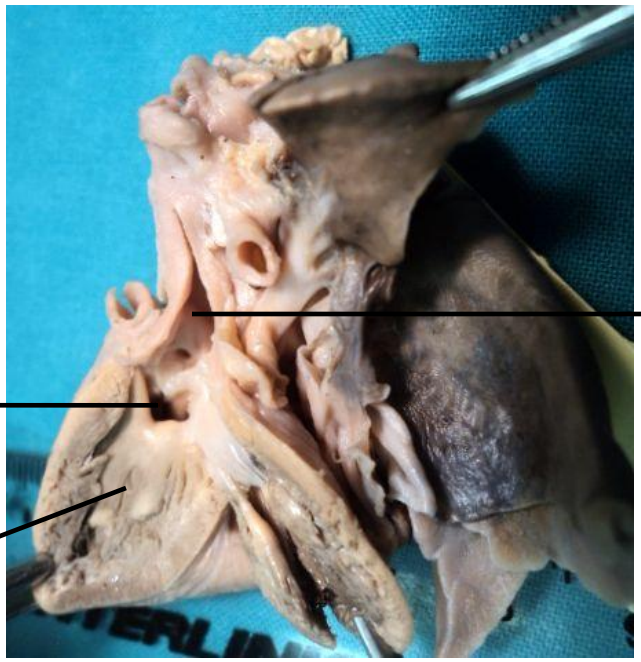


Figure 3.12: Case 5- Post-mortem specimen with tetralogy of Fallot displaying a large perimembranous ventricular septal defect [Photographed from Professor Levin's post mortem collection]

In case six, the child was diagnosed with a severe case of tetralogy of Fallot. The large ascending aorta (26 millimeters) was displaced anteriorly with marked override (Fig. 3.13). The main pulmonary artery trunk (11 millimeters) was observed beside the large aorta. The right ventricle was enlarged and hypertrophied (Fig. 3.13). The perimembranous ventricular septal defect measured 14 millimeters (Fig. 3.14). Narrowing of the right ventricular outflow tract was observed.

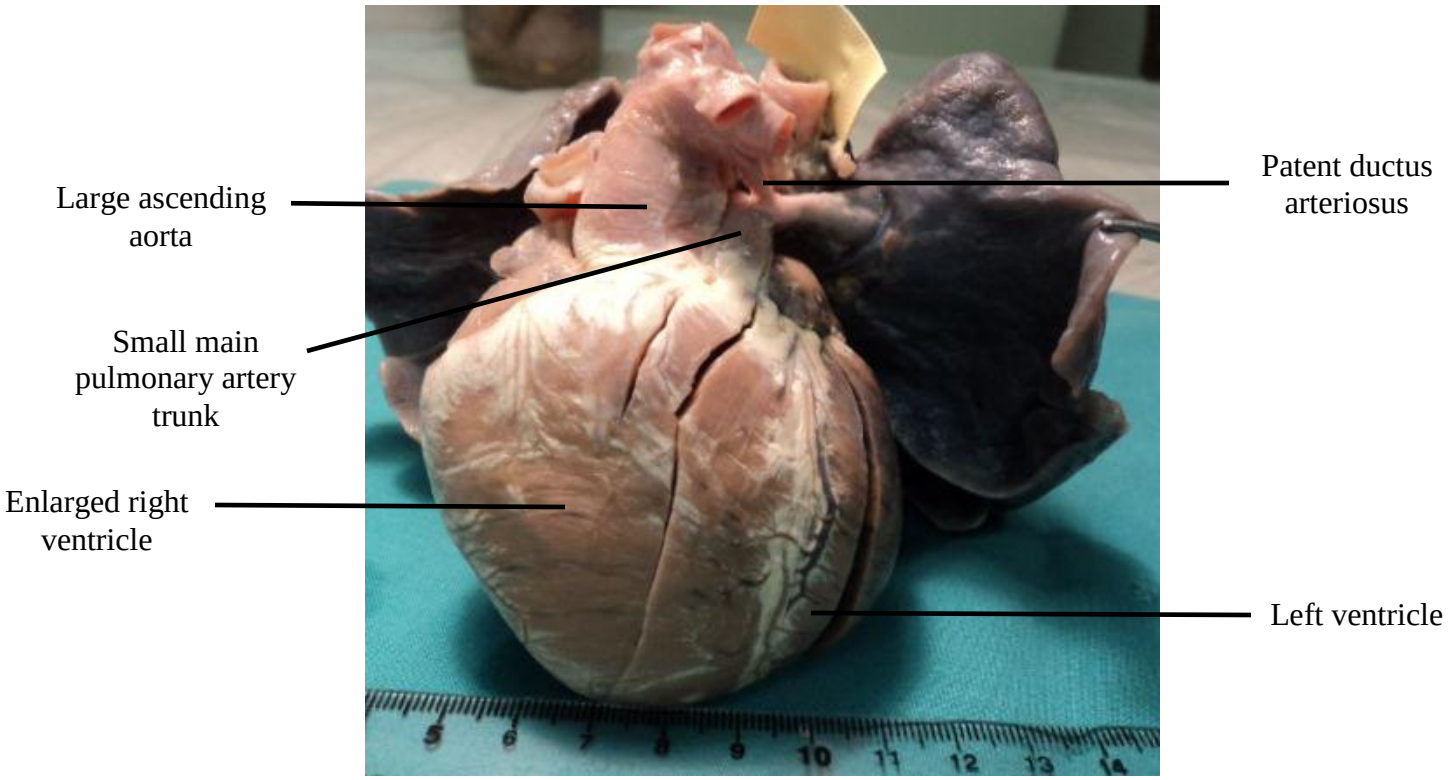


Figure 3.13: Case 6- Frontal view of a post-mortem specimen of a case diagnosed with tetralogy of Fallot demonstrating a large aorta and enlarged right ventricle
[Photographed from Professor Levin's post mortem collection]

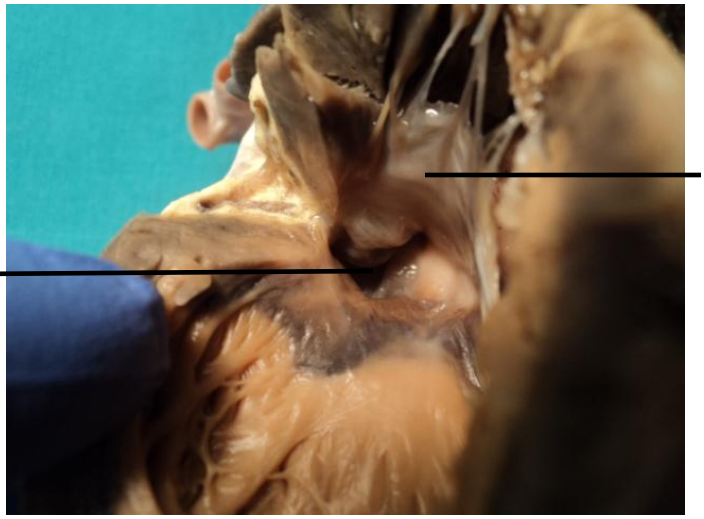


Figure 3.14: Case 6- Post-mortem specimen of a case diagnosed with tetralogy of Fallot demonstrating the large perimembranous ventricular septal defect from the left ventricle
[Photographed from Professor Levin's post mortem collection]

In case seven, no clinical information was available on record. The examination of the post-mortem specimen revealed a large aorta (12 millimeters) and a small main pulmonary artery trunk (six millimeters) (Fig. 3.15). The right ventricular outflow tract was narrowed and thickened and the right ventricle was severely hypertrophied (Fig. 3.15). The perimembranous ventricular septal defect measured five millimeters (Fig. 3. 16).

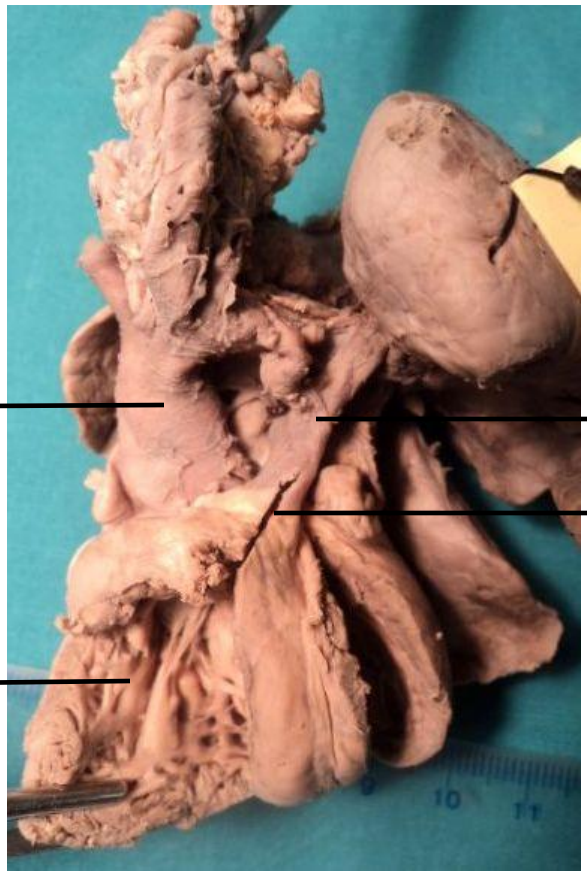


Figure 3.15: Case 7- Post-mortem specimen of a case diagnosed with tetralogy of Fallot demonstrating the large aorta, hypertrophied right ventricle and the right ventricular outflow tract
[Photographed from Professor Levin's post mortem collection]

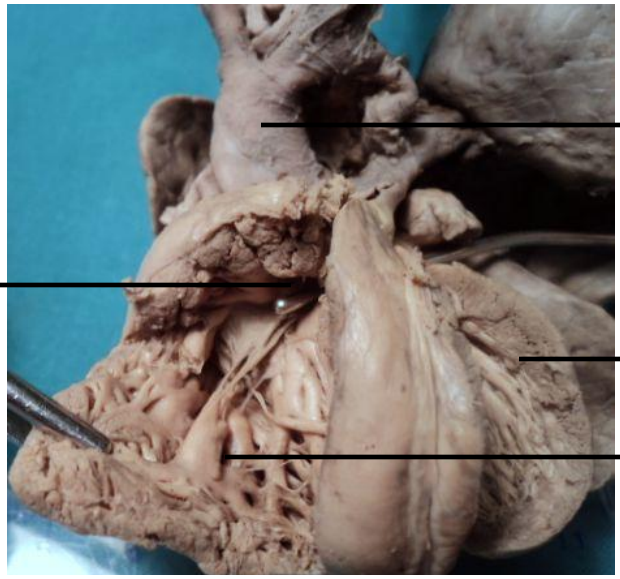


Figure 3.16: Case 7- Post-mortem specimen of a case diagnosed with tetralogy of Fallot demonstrating the perimembranous ventricular septal defect from the right ventricle
[Photographed from Professor Levin's post mortem collection]

3.2 Retrospective study of clinical records

3.2.1 Ventricular septal defects as an isolated defect

Forty-seven of the 50 infant records which were analysed were born by normal vaginal delivery. Three of the remaining infants experienced complicated births. Two of the latter three were premature and were delivered by Caesarian section. The remaining one was also delivered by Caesarian section due to fetal distress.

There were more females than males in the total sample (females n = 30; males n = 20). The predominant population group was Black (n = 43). Of the remainder, there were three White, three Indian and one Coloured child in the sample. The race of the children was confirmed directly from the surgeon who operated on them.

The age distribution of the total sample (n=50) ranged from 4 months to 13 years with a mean age of 3 years (standard deviation = ± 3 years) (Table 3.3). Twenty cases were found to be below 2 years of age and presented with a mean age of 9 months. In the 2-4 year age group (n = 17), the mean age was 2 years 6 months. Thirteen cases were over the age of 4 years with the minimum age being 5 years and maximum being 13 years of age (Table 3.3).

The weight and height of the entire sample ranged from 3.3 to 31 kg (mean = 10.5 kg, standard deviation = ± 5.4 kg) and from 0.5 to 1.3 m (mean = 0.8 m, standard deviation = ± 0.2m) respectively. In three cases, heights were not recorded in the patient reports.

First presentation of the ventricular septal defect and the details of the subsequent operation

In group A (n=20), a ventricular septal defect was already diagnosed at a pre-hospital admission in three of the cases. In another two cases, the children were referred to the cardiac clinic because of respiratory distress (Table 3.4). The latter babies presented with a history of tachypnoea, shortness of breath and a cough. A cardiac murmur was heard at some time after birth in eleven of the cases before their ventricular septal defect was diagnosed.

In group B (n=17), three of the cases where referred to the cardiac clinic with a diagnosed ventricular septal defect, while eight of the cases were referred because of a cardiac murmur (Table 3.5). In addition, another three of the cases were referred to the cardiac clinic because the infants were in congestive cardiac failure.

In group C (n=13), five of the cases were referred to the cardiac clinic with a diagnosis of ventricular septal defect (Table 3.6). Four of the cases were referred with a history of swelling of the feet, dyspnoea and a recurrent cough. A cardiac murmur was heard in four cases (Table 3.6).

All the initial diagnoses shown in the tables above resulted in further investigations following which an accurate diagnosis of the ventricular septal defect was made.

Due to the increased workload of surgeons and the lack of sufficient resources at government hospitals over the period assessed, in most instances the cases diagnosed with a ventricular septal

defect did not undergo operation immediately. Forty-four of the 50 cases were placed on a waiting list.

In one of the remaining six cases, the surgeons felt that the ventricular septal defect was very small and would close spontaneously. In another case, the infant was under weight, and the surgeons felt that it would be best to wait for the child to gain weight prior to the operation. Two of the remaining cases were considered high risk for surgery due to severe pulmonary hypertension. In a further case the ventricular septal defect was almost closed by the prolapse of the right coronary cusp of the aortic valve into the ventricular septal defect. In the remaining case, the ventricular septal defect was not closed immediately, but underwent palliative surgery which included banding of the main pulmonary artery and repair of the coarctation of the aorta.

Additional cardiac operations prior to ventricular septal defect repair

In four of the 50 cases, the patients underwent a cardiac operation prior to the surgery for repair of the ventricular septal defect. In one of these, a patent ductus arteriosus was ligated and no complications were noted post-operatively. In another case, the child underwent a main pulmonary artery banding. This operation was abandoned, as the extubation failed on five occasions.

In a further case, several previous operative procedures had been carried out. The infant underwent an operation for closure of a 12 mm perimembranous ventricular septal defect, a patent ductus arteriosus ligation and excision of the atrial septum with re- routing of the left

superior vena cava into the right atrium using a bovine pericardial patch. Post-operatively, several complications ensued viz. high pulmonary artery and left atrial pressures, while the coronary sinus patch protruded into the left atrium. Pulmonary hypertension occurred due to a residual ventricular septal defect. Pulmonary venous obstruction was caused by the atrial septal defect patch. This child was taken back to theatre for closure of an additional ventricular septal defect (inlet). This patient had been included in this series because of surgery for the second residual ventricular septal defect.

In the final case, the infant underwent repair of a coarctation of the aorta (with a subclavian flap) as well as pulmonary artery banding. Dense adhesions formed in the region of the band. The band was very tight and impinged on the right pulmonary artery.

Echocardiographic diagnosis of the type and site of ventricular septal defect

Forty-five (90%) of the 50 cases had a single ventricular septal defect. Of these, a perimembranous ventricular septal defect was the most common type diagnosed (n= 35; 78%) (Fig.3.17). Six muscular ventricular septal defects were identified (13%); three infants with outlet ventricular septal defects (7%), and only one child had an inlet ventricular septal defect (2%).

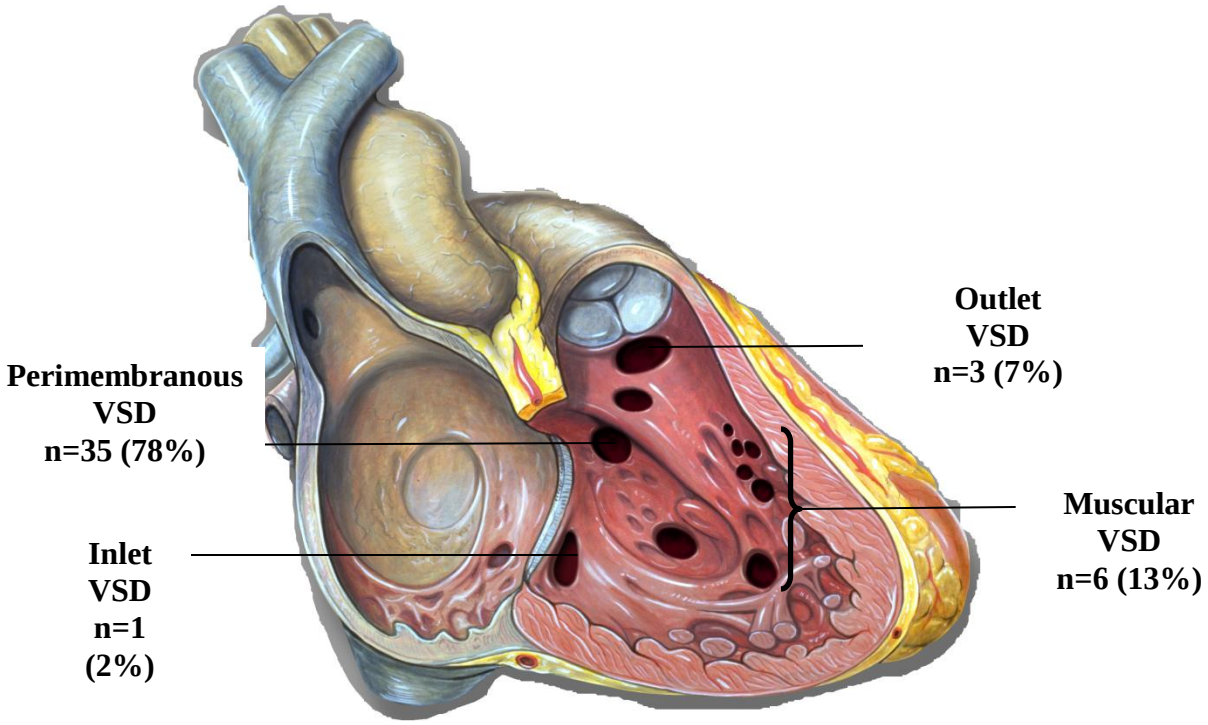


Figure 3.17: Right ventricular view of the interior of the heart indicating the number and location of the different types of single ventricular septal defects identified in the present retrospective study
(Adapted from: http://commons.wikimedia.org/wiki/File:Heart_right_vsd.jpg

Multiple ventricular septal defects were found in five of the 50 patients (10%). All five had perimembranous ventricular septal defects. In addition, two had apical muscular ventricular septal defects (Fig.3.18). A mid-muscular ventricular septal defect was diagnosed in the third case, and another had two mid-muscular ventricular septal defects. In the final case, there was a mid-muscular and Swiss cheese defect (multiple small ventricular septal defects) (Fig.3.18).

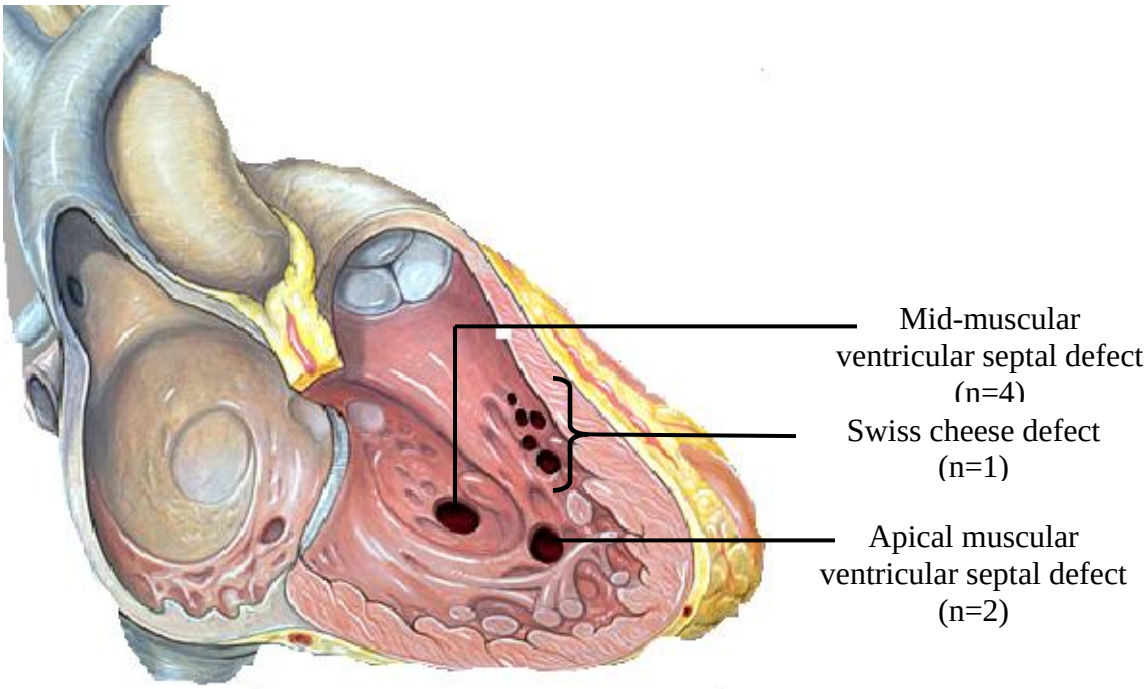


Figure 3.18: Right ventricular view of the interior of the heart indicating the number and location of the different types of multiple ventricular septal defects identified in the present retrospective study
(Adapted from: http://commons.wikimedia.org/wiki/File:Heart_right_vsd.jpg)

Haemodynamic consequence of the ventricular septal defect

Shunting of blood was assessed by Colour Doppler Echocardiography and was left to right in 37 of the 45 cases with a single ventricular septal defect (Table 3.7). Six of the cases were found to have bi-directional shunting. In the remaining two patients (one perimembranous and one inlet), there was no documentation of the shunting.

For the cases with multiple ventricular septal defects, in three of the five cases, there was left to right shunting through the ventricular septal defect. In the remaining two cases, bi-directional shunting was documented.

Diagnosis of additional cardiac abnormalities

In group A (younger than two years), 15 of the 20 cases presented with other cardiac defects in addition to, or as a result of the ventricular septal defect. Two cases showed prolapse of the right coronary cusp into the ventricular septal defect (one perimembranous and one outlet defect), diagnosed by echocardiography. One of these cases, with the perimembranous defect, had mild aortic regurgitation. Seven cases had a patent ductus arteriosus (2-4mm in diameter) and all of these ductuse were ligated during the operation for the ventricular septal defect. Four cases presented with atrial septal defects (two small: 4 and 5mm and two moderate in size: 8 and 9mm) (Table 3.8). These defects were also closed during the operation for the ventricular septal defect.

Additional abnormalities that were documented included, cor triatriatum (suspicion of membrane in left atrium proximal to atrial septal defect) in one case, and interrupted inferior vena cava with azygous continuation in another case (Table 3.8).

In group B (2-4 years), additional cardiac abnormalities were present in 13 out of the 17 patients (Table 3.9). Prolapse of the right coronary cusp into the perimembranous ventricular septal defect was documented in three patients and aortic regurgitation occurred in three patients (two of whom had prolapse of the right coronary cusp). A discrete subaortic stenosis occurred in five cases, four had a patent ductus arteriosus (2-3mm), and one further case presented with a large atrial septal defect (19mm in diameter) (Table 3.9). This defect was closed during the operation for the ventricular septal defect.

In group C (over four years), additional cardiac abnormalities were diagnosed in 12 of the 13 patients (Table 3.10). Six cases had discrete subaortic stenosis and six cases had prolapse of the right coronary cusp into the ventricular septal defect (five perimembranous defects and one outlet defect). Two of the cases with perimembranous ventricular septal defects and aortic valve prolapse, also had aortic regurgitation. Three cases had a patent ductus arteriosus (3-4mm) and only one case presented with an atrial septal defect (12mm) (Table 3.10). These defects were closed during the operation for the ventricular septal defect.

Anatomical location and size of the ventricular septal defects

In the cases where only a single ventricular septal defect was found (n = 45), 35 had a perimembranous ventricular septal defect (20 were described in the echocardiographic reports as large, seven were moderate and eight were small in size) (Table 3.11). Of the remaining 10 cases, six muscular ventricular septal defects, three outlet ventricular septal defects and one inlet ventricular septal defect were seen.

The size of the ventricular septal defect was also assessed from the measurement of two parameters viz. the ratio of ventricular septal defect diameter to aortic annulus diameter as measured on echocardiography, and the degree of systolic pulmonary hypertension as recorded at cardiac catheterization. If the ventricular septal defect to aortic annulus ratio was greater than 50% and if the systolic pulmonary artery pressure was greater than 40mmHg, the ventricular septal defect and the shunt across the defect was regarded as large. The raw data for the systolic

pulmonary artery pressures appears in Appendix A. The aortic annulus diameter was only recorded in 19 patients; therefore these ratios were not used in this study.

The average size of a large perimembranous ventricular septal defect was 11.7 millimeters when measured from the echocardiogram (only 19 of the 20 measurements were recorded) (Table 3.11). The average size of the moderate perimembranous ventricular septal defect measured 7.6 millimeters from the echocardiogram (measurements were recorded in only 4 of the 7 cases). The average size of the small perimembranous ventricular septal defect from the echocardiogram measured 5.8 millimeters.

Analysis of type and size of ventricular septal defects by age groups

In group A (n= 16) (younger than two years) 12 of the infants were diagnosed with perimembranous ventricular septal defects, (large: n = 8 and small: n= 4) (Table 3.12). One infant each was diagnosed as having an outlet and an inlet ventricular septal defect while two children had muscular defects. The average size of a large perimembranous ventricular septal defect was found to be 9.9 millimeters from the echocardiogram (Table 3.12).

In group B (2-4 years), 14 out of the 17 infants were diagnosed with perimembranous ventricular septal defects. Six of these were large (average size was 10.9 millimeters when measured from the echocardiogram) (Table 3.13). The average size of the six moderate perimembranous ventricular septal defects was 7.1 millimeters from the echocardiogram (only 4 of the 5 measurements were recorded). Three small perimembranous ventricular septal defects, two muscular ventricular septal defects and one outlet ventricular septal defect were diagnosed

(Table 3.13). The two muscular defects were regarded as moderate in size in the echocardiographic reports, while the single outlet ventricular septal defect was large.
In group C (over 4 years of age), nine out of the 12 infants were diagnosed with perimembranous ventricular septal defects, (large: n = 6; moderate: n = 2 and small: n= 1) (Table 3.14). The average size of the large perimembranous ventricular septal defect was 15.4 millimeters when recorded from the echocardiogram (only 5 of the 6 measurements were recorded) (Table 3.14). Two muscular and one outlet ventricular septal defect were diagnosed. None of the infants had an inlet ventricular septal defect.

In the cases with multiple ventricular septal defects, all five of the infants had a perimembranous ventricular septal defect, and in addition a muscular ventricular septal defect (apical muscular in one case and mid-muscular in four cases) (Table 3.15). In one case, that of a one year old Black female, Swiss cheese defects were diagnosed in addition to the perimembranous and mid-muscular ventricular septal defects. In a further one case, two mid-muscular ventricular septal defects were diagnosed in addition to the perimembranous ventricular septal defect.

As shown in Table 3.15 many of the measurements were not recorded in the echocardiographic reports. However all the measurements for the perimembranous ventricular septal defect were recorded. The size of the smaller muscular ventricular septal defects could possibly have made the measuring of the ventricular septal defects difficult and this may have contributed to the absence of the data.

3.2.2 Ventricular septal defects and the site of right ventricular outflow tract obstruction in tetralogy of Fallot

A total of 42 infants were diagnosed as having tetralogy of Fallot over the period of four years (2001 to 2004). Forty of the 42 infants were born by normal vaginal delivery. Two of the remaining infants experienced complicated births. One of the two was premature, while the other was delivered by Caesarian section due to fetal distress.

The age distribution of these cases ranged from 1 year 2 months to 13 years with a mean age of 3 years 5 months (Table 3.16). Thirteen cases were found to be below 2 years of age and presented at a mean age of 1 year 4 months. In the 2-4 year age group (n = 16), the mean age was 2 years 7 months. Thirteen cases were over 4 years with the minimum age being 5 years and maximum being 13 years of age.

There were equal numbers of males and females in the total sample (males n = 21; females n = 21). The predominant population group was Black (n = 33 out of 42) (78.6%). There were three White, five Coloured children, and one Indian child in the rest of the sample.

Of the 42 patients, the weight (n=40) ranged from 6.4 to 21.4 kg (mean = 11.7 kg and standard deviation = ± 3.9kg) and the height in 38 cases from 0.6 to 1.50 m (mean = 0.9 m and standard deviation = ± 0.2m). In two cases, weight, and in four cases, height, were not recorded.

The type of ventricular septal defect and the site of right ventricular outflow tract obstruction in tetralogy of Fallot

A perimembranous ventricular septal defect was the most common site identified in patients with tetralogy of Fallot (n= 38; 90%). The remaining four cases (10%) all had an outlet type of ventricular septal defect (Fig.3.19).

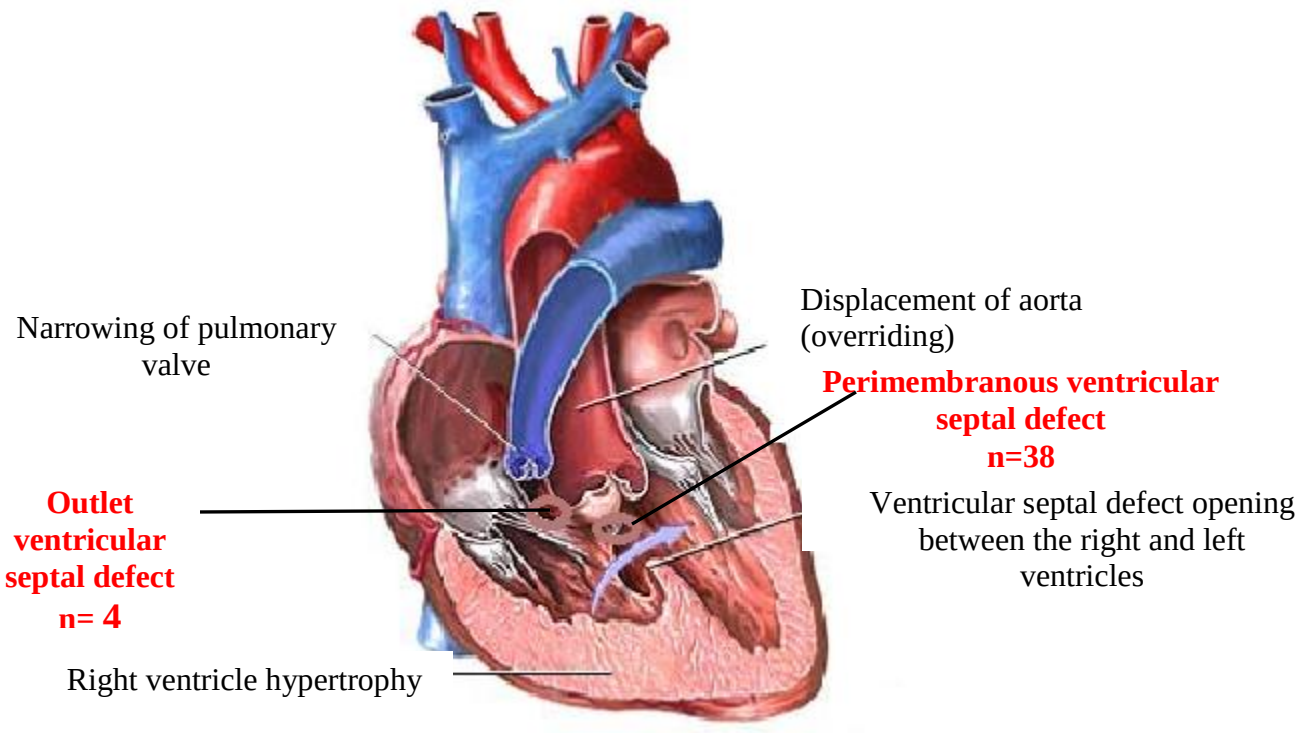


Figure 3.19: Coronal section of the heart indicating the number and location of the different types of ventricular septal defects in tetralogy of Fallot identified in the present retrospective study
(Adapted from: http://www.doctortipster.com/3146-tetralogy-of-Fallot-symptoms-diagnosis-and-treatment.html)

Haemodynamic consequence of the ventricular septal defects in tetralogy of Fallot

Fifteen cases of the perimembranous ventricular septal defects in tetralogy of Fallot presented with right to left shunting of blood, fourteen cases with bi-directional shunting and in four cases left to right shunting was observed. In the remaining five cases, the haemodynamic consequence of the ventricular septal defect was not documented. For the outlet ventricular septal defects, two of the four cases were noted to have right to left shunting, one case with left to right shunting and the remaining one case with bi-directional shunting of blood.

Diagnosis of the site of right ventricular outflow tract obstruction and additional cardiac abnormalities occurring with tetralogy of Fallot

In group A (younger than 2 years) (n=13), infundibular and pulmonary valve stenosis was found in all thirteen cases. Two cases in addition, had a small patent ductus arteriosus and two other cases had small atrial septal defects (Table 3.17).

In group B (2-4 years) (n=16), infundibular and/or pulmonary valve stenosis were observed in 13 and eight cases respectively. Additional cardiac abnormalities were present in five cases, two with a small patent ductus arteriosus and one a small atrial septal defect (Table 3.18). Discrete subaortic stenosis was found in one case, and in another a double outlet right ventricle was observed.

In group C (older than 4 years) (n=13), infundibular and/or pulmonary valve stenosis was observed in nine and six cases respectively. In addition, two patients had small atrial septal defects and one had a small patent ductus arteriosus (Table 3.19).

First presentation of tetralogy of Fallot and the subsequent operation details

In group A (younger than 2 years) (n=13), three of the cases were referred to the cardiac clinic with a diagnosis of tetralogy of Fallot. Five of the cases were referred because of respiratory distress (Table 3.20). They presented with a history of severe cyanotic spells, shortness of breath and a cough. A cardiac murmur was heard at birth in five of the cases.

In group B (2-4 years) (n=16), seven of the cases were referred with a diagnosis of tetralogy of Fallot (Table 3.21). Three of the cases were referred with a cardiac murmur, while five of the cases had a history of respiratory distress due to severe and frequent cyanotic spells.

In group C (older than 4 years) (n=13), ten of the cases were referred with a diagnosis of tetralogy of Fallot (Table 3.22). Two of the cases were referred with a history of respiratory distress and cyanosis. In one case, the referral was made for a cardiac murmur (Table 3.22).

Anatomical location and size of the ventricular septal defects in tetralogy of Fallot

The ventricular septal defect was found in the perimembranous position in 38 (90%) of the 42 cases (all assessed as large in the echocardiographic reports), and in the outlet region in four children (10%) (Table 3.23). The average size of the perimembranous ventricular septal defect was 10.4mm as measured from the echocardiogram (only 11 of the 38 measurements were recorded) (Table 3.23). The average size of the four outlet ventricular septal defects measured 10mm from the echocardiogram.

In the young group (under two years of age) (n=13), ten of the infants were diagnosed with large perimembranous ventricular septal defects and three were diagnosed with outlet ventricular septal defects (the size of ventricular septal defects that were recorded appear in Table 3.23).

In group B (2-4 years) all 16 of the infants had large perimembranous ventricular septal defects (measurements recorded in four of 16 cases).

In group C (older than 4 years) (n=13), 12 infants had large perimembranous ventricular septal defects and one outlet ventricular septal defect was diagnosed (Table 3.23).

Once again due to the increased workload of surgeons and the lack of sufficient resources at the Johannesburg hospital, in most instances, the infants diagnosed with tetralogy of Fallot were not operated on immediately. Forty cases where placed on the waiting list for an operation date. In two cases, a Blalock-Taussig shunt was carried out urgently as a palliative procedure prior to surgical correction for tetralogy of Fallot.

3.2.3 Ventricular septal defects and right ventricular outflow tract obstruction in double chambered right ventricle

Eight infants were identified with a double chambered right ventricle. All eight of the infants were born by normal vaginal delivery.

The age distribution of this sample ranged from 6 months to 6 years with a mean age of 2 years 7 months (Table 3.24). In group A, there were two cases, one at age 6 months and the other at 8

months. In group B (n = 5), the mean age was 2 years 8 months. There was only one child in group C (6 years old).

In this small sample (n=8), there were more males than females (males n = 6; females n = 2). Only the Black population group was represented in the sample (n = 8).

The weight and height of seven of the eight patients ranged from 6.9 to 12.7 kg (mean = 10.2 kg and standard deviation = ± 2.1 kg) and from 0.6 to 0.9 m (mean = 0.8 m) respectively. In one case both weight and height were not recorded.

Diagnosis of the type of ventricular septal defect occurring with the double chambered right ventricle

Perimembranous ventricular septal defect was the most common type of ventricular septal defect diagnosed in this group (n= 6; 75%). The remaining two cases (25%) had an outlet type of ventricular septal defect.

Haemodynamic consequence of the ventricular septal defect in double chambered right ventricle

Five cases of the perimembranous ventricular septal defect presented with left to right shunting of blood, and one case with bi-directional shunting was observed. For the outlet ventricular septal defects, one of the two cases was diagnosed as having right to left shunting and the remaining one case with left to right shunting of blood.

Diagnosis of right ventricular site of obstruction and additional cardiac abnormalities

In all eight of the cases in this group, a large hypertrophied parietal muscle bundle was seen in the outflow area of the right ventricle which is the characteristic feature of double chambered right ventricle. These muscle bundles were resected during the surgery for closure of the ventricular septal defects.

In group A (younger than 2 years), one case had a small patent ductus arteriosus.

In group B (2-4 years), discrete subaortic stenosis was present in two of the five patients.

In group C (6 years old), this single patient also had discrete subaortic stenosis.

First clinical presentation of double chambered right ventricle and the subsequent operation details

In group A (younger than 2 years) (n=2), one of the cases was referred to the cardiac clinic for a cardiac murmur heard at birth, and the other due to bronchopneumonia.

In group B (2-4 years) (n=5), one case was referred to the cardiac clinic with the diagnosis of double chambered right ventricle. Four of the cases were referred to because of a cardiac murmur.

In group C (6 years old), this case was referred with signs of congestive cardiac failure.

Seven cases were placed on the waiting list for an operation date. In the remaining one case, the insertion of a Blalock-Taussig shunt as a palliative procedure was considered more urgent because of severe cyanosis.

Type and size of the ventricular septal defect as documented by the cardiologist from the echocardiogram

In the total sample (n=8) with double chambered right ventricle, six of the infants were diagnosed with perimembranous ventricular septal defects, of which three were moderate and three were small in size (Table 3.25). Two had outlet ventricular septal defects.

The average size of the three moderate perimembranous ventricular septal defects measured 7.3 mm on the echocardiogram. The average size of the small perimembranous ventricular septal defects was found to be 3.8 mm from the echocardiogram (only two of the three measurements were recorded) (Table 3.25). Only one of the two outlet ventricular septal defect measurements was recorded.

In group A (younger than 2 years), in one of the two infants there was a moderate perimembranous ventricular septal defect (9 mm on echocardiogram). The other infant had an outlet ventricular septal defect but no measurement was recorded.

In group B (2-4 years), all five of the infants were diagnosed with perimembranous ventricular septal defects, (moderate: n = 2 and small: n= 3). The average size of the moderate perimembranous ventricular septal defects was 9.7 mm from the echocardiogram. The average size of two of the three small perimembranous ventricular septal defects from the echocardiogram was 3.8 mm. There was no record in the third patient.

In group C, the one infant (6 years old) was diagnosed with an outlet ventricular septal defect which measured 11 mm from the echocardiogram.

SUMMARY OF RESULTS

In this retrospective clinical study, a perimembranous ventricular septal defect was the most common type of ventricular septal defect observed between the years 2001 to 2004 in Johannesburg provincial hospitals. Perimembranous ventricular septal defects were found in 78% (n= 35) of the 45 cases that had a single isolated ventricular septal defect, in 90% (n=38) of the 42 patients diagnosed with tetralogy of Fallot, and in 75% (n=6) of the eight patients in the group with double chambered right ventricle.

An outlet type of ventricular septal defect was the second most common type of ventricular septal defect found in this study. The outlet ventricular septal defect was found in three infants (7%) out of the 45 cases with isolated ventricular septal defects, in four cases (10%) out of 42 patients with tetralogy of Fallot and in two (25%) of the eight cases with double chambered right ventricle.

In this entire study, the muscular and inlet type of ventricular septal defect were found exclusively in the group of ventricular septal defects as isolated defects (13% and 2% of the 45 cases respectively).

TABLES

Table 3.1: Calculated ages of the 11 neonatal cadavers from the School of Anatomical Sciences using foot-length, crown-rump length and bi-parietal breadth

	Sex	*Race*	*Mean foot length (mm)*	*Mean crown rump length (mm)*	*Mean bi-parietal breadth (mm)*	*Calculated age (weeks) (Moore & Persaud, 1998)*
1.	**F**	**Black**	**82.0**	**37.2**	**79.8**	**38-40**
2.	**M**	**Black**	**81.5**	**34.1**	**85.3**	**36-38**
3.	**F**	**Black**	**61.6**	**31.4**	**76.3**	**30-32**
4.	**M**	**Black**	**62.5**	**31.9**	**84.1**	**30-32**
5.	**F**	**Black**	**60.6**	**32.5**	**86.4**	**28-30**
6.	**F**	**Black**	**53.8**	**30.0**	**65.9**	**28-30**
7.	**F**	**Black**	**56.0**	**30.9**	**73.3**	**28-30**
8.	**M**	**Black**	**55.7**	**30.4**	**77.5**	**28-30**
9.	**F**	**Black**	**52.2**	**30.7**	**80.6**	**28-30**
10.	**M**	**Black**	**53.6**	**29.6**	**78.4**	**26-28**
11.	**F**	**Black**	**50.3**	**26.4**	**66.7**	**24-26**

Table 3.2: Mean length and breadth of the different components of the interventricular septum in eight neonatal cadavers measured in millimeters (mm)

Neonate No	Mean length (mm)				Mean breadth (mm)		Age in weeks
	Inlet Septum	Outlet Septum	Muscular Septum	Perimembranous Septum	Muscular Septum	Perimembranous Septum	
1.	**9.6**	**5.4**	**17.2**	**12.1**	**15.9**	**23.3**	**38-40**
2.	**6.9**	**4.7**	**18.2**	**4.0**	**15.9**	**16.6**	**30-32**
3.	**5.5**	**3.4**	**16.5**	**8.4**	**18.1**	**22.9**	**30-32**
4.	**3.6**	**5.4**	**16.0**	**8.7**	**19.0**	**18.6**	**28-30**
5.	**5.3**	**3.3**	**16.5**	**6.7**	**17.6**	**17.4**	**28-30**
6.	**7.4**	**8.0**	**16.8**	**9.8**	**18.1**	**23.5**	**28-30**
7.	**8.0**	**5.2**	**16.0**	**6.8**	**10.8**	**21.7**	**28-30**
8.	**4.5**	**4.1**	**18.0**	**5.7**	**15.9**	**19.5**	**24-26**

Table 3.3: Age distribution and grouping of the cases with an isolated ventricular septal defect

Age	*Number*	*Minimum*	*Maximum*	*Mean*	*Standard Deviation*
Total sample	50	4 months	13 years	3 years 1 month	± 3 years
A) Younger than 2 years	20	4 months	1 year 6 months	9 months	± 3 months
B) 2 - 4 years	17	2 years	4 years	2 years 6 months	± 8 months
C) Older than 4 years	13	5 years	13 years	12 years 1 month	± 2 years 7 months

Table 3.4: Group A (younger than two years): First clinical presentation of the ventricular septal defect (VSD)

Reason for initial VSD diagnoses	*Number of cases*
Cardiac murmur	11
Bronchopneumonia	4
VSD already diagnosed	3
Respiratory distress	2
Total	**20**

Table 3.5: Group B (2-4 years): First clinical presentation of the ventricular septal defect

Reason for initial VSD diagnoses	*Number of cases*
Cardiac murmur	8
VSD already diagnosed	3
Recurrent respiratory tract infections	3
Congestive cardiac failure	3
Total	**17**

Table 3.6: Group C (older than 4 years): First clinical presentation of the ventricular septal defect

Reason for initial VSD diagnoses	*Number of cases*
VSD already diagnosed	5
Presented with history of swelling of feet, dyspnoea & cough	4
Cardiac murmur	4
Total	**13**

Table 3.7: Haemodynamic consequence of a single ventricular septal defect (VSD)

Isolated VSD (n=45)	***Left to right shunting***	***Bi-directional shunting***	***Shunting not recorded***
Perimembranous VSD			
Total sample (n=35)	30	4	1
Muscular VSD			
Total sample (n=6)	4	2	-
Outlet VSD			
Total sample (n=3)	3	-	-
Inlet VSD			
Total sample (n=1)	-	-	1

Table 3.8: Group A (younger than 2 years): Additional cardiac abnormalities associated with the ventricular septal defect (VSD)

No	Age	Sex	Race	Prolapse of RCC into VSD	PDA	ASD	Other
1.	4 months	M	B	×	3mm	×	Thickened Interatrial septum
2.	5 months	M	B	×	×	4mm	×
3.	5 months	M	I	×	3mm	×	Interrupted inferior vena cava with azygos continuation
4.	5 months	F	W	×	2mm	9mm	Cor triatriatum
5.	7 months	F	B	×	×	5mm	×
6.	7 months	F	B	×	×	×	×
7.	8 months	F	B	√	×	×	×
8.	1 year	M	W	×	×	8mm	×
9.	1 year	M	B	×	×	×	×
10.	1 year	M	W	×	3mm	×	Mesocardia (partial rotation of the heart to the right)
11.	1 year	F	B	×	4mm	×	×
12.	1 year	F	B	×	×	×	×
13.	1 year	M	I	×	4mm	×	×
14.	1 year 5 months	F	B	×	√	×	×
15.	1 year 6 months	M	B	√	×	×	Mild aortic regurgitation. Bilateral superior vena cavae

RCC=right coronary cusp; PDA=patent ductus arteriosus; ASD=atrial septal defect
× = absent; √ = present

Table 3.9: Group B (2-4 years): Additional cardiac abnormalities associated with the ventricular septal defect (VSD)

No	Age	Sex	Race	Prolapse of RCC into VSD	PDA	ASD	DSAS	AR	Other
1.	2	F	B	×	×	×	√	×	Posterior malaligned aorta
2.	2	F	B	×	×	×	×	×	Minor aneurysm of right sinus of Valsalva
3.	2	F	B	×	×	19mm	×	×	Bilateral superior vena cavae. Absent innominate vein
4.	2	M	B	√	×	×	×	×	×
5.	2	M	B	×	3mm	×	×	×	×
6.	2	M	B	√	×	×	√	√	×
7.	2	F	B	×	×	×	×	×	Left pulmonary artery stenosis
8.	2	M	B	×	×	×	√	×	×
9.	3	F	B	×	2mm	×	×	×	×
10.	3	M	B	×	3mm	×	√	×	×
11.	4	F	B	×	3mm	×	×	×	Double outlet right ventricle
12.	4	M	B	×	×	×	×	√	Non discrete left ventricular outflow tract obstruction
13.	4	F	B	√	×	×	√	√	×

RCC=right coronary cusp; PDA=patent ductus arteriosus; ASD=atrial septal defect;
DSAS=discrete subaortic stenosis; AR=aortic regurgitation
× = absent; √ = present

Table 3.10: Group C (over 4 years): Additional cardiac abnormalities associated with the ventricular septal defect (VSD)

No	Age	Sex	Race	Prolapse of RCC into VSD	PDA	ASD	DSAS	AR	Other
1.	5	M	B	√	×	×	√	×	×
2.	5	M	B	×	3mm	×	×	×	×
3.	5	M	B	√	3mm	×	√	√	×
4.	6	F	B	×	4mm	×	×	×	×
5.	6	F	B	×	×	12mm	×	×	×
6.	7	F	B	√	×	×	×	×	×
7.	7	F	B	×	×	×	√	×	Bilateral superior vena cavae. Absent innominate vein
8.	7	F	B	×	×	×	√	×	Double outlet right ventricle
9.	7	F	B	√	×	×	√	×	×
10.	10	F	C	√	×	×	√	×	×
11.	12	F	B	×	×	×	×	×	×
12.	13	F	B	√	×	×	×	√	×

RCC=right coronary cusp; PDA=patent ductus arteriosus; ASD=atrial septal defect;
DSAS=discrete subaortic stenosis; AR=aortic regurgitation
× = absent; √ = present

Table 3.11: Type and average size of the single ventricular septal defects (VSD) documented from the echocardiographic reports (n=45)

	Perimembranous VSD n=35			Muscular n=6	Outlet n=3	Inlet n=1
	Large n=20 (19)	*Moderate* n=7 (4)	*Small* n=8 (8)			
Echocardiogram	**11.7mm**	**7.6mm**	**5.8mm**	**9.3mm**	**10.3mm**	**9.0mm**

N.B. Figures in brackets are the number of infants in which the size of the VSD was recorded

Table 3.12: Type and average size of the ventricular septal defect (VSD) documented from the echocardiographic reports in the cases with single VSDs in group A (younger than 2 years)

	Perimembranous VSD n=12		Muscular n=2	Outlet n=1	Inlet n=1
	Large n=8	*Small* n=4			
Echocardiogram	**9.9mm**	**6.0mm**	**9.0mm**	**7.0mm**	**9.0mm**

Table 3.13: Type and average size of the ventricular septal defect (VSD) documented from the echocardiographic reports in the cases with single VSDs in group B (2-4 years)

	Perimembranous VSD n=14			**Muscular n=2**	**Outlet n=1**
	Large **n=6 (6)**	*Moderate* **n=5 (4)**	*Small* **n=3 (3)**		
Echocardiogram	**10.9mm**	**7.1mm**	**6.8mm**	**8.5mm**	**14.0mm**

N.B. Figures in brackets are the number of infants in which the size of the VSD was recorded

Table 3.14: Type and average size of the ventricular septal defect (VSD) documented from the echocardiographic reports in the cases with single VSDs in group C (over 4 years of age)

	Perimembranous VSD n=9			**Muscular n=2**	**Outlet n=1**
	Large **n=6 (5)**	*Moderate* **n=2 (1)**	*Small* **n=1**		
Echocardiogram	**15.4mm**	**6.0mm**	**5.0mm**	**10.5mm**	**10.0mm**

N.B. Figures in brackets are the number of infants in which the size of the VSD was recorded

Table 3.15: Type and size of the ventricular septal defect documented from the echocardiographic reports in the cases with multiple ventricular septal defects

No.	Age	Sex	Race	Type of ventricular septal defect	Size of ventricular septal defect (mm)	
					Echocardiogram	
					PM	M
1.	6 months	F	White	Perimembranous and apical muscular	4	-
2.	1 year	F	Black	Perimembranous and mid-muscular	10	6
3.	1 year	F	Black	Perimembranous, mid-muscular & Swiss cheese	-	-
4.	1 year 5 months	F	Black	Perimembranous and mid-muscular	-	7
5.	12 years	F	Black	Perimembranous and 2 mid-muscular	8.5	-

PM= perimembranous; M= muscular

Table 3.16: Age distribution and groupings of the infants with tetralogy of Fallot

Age (yrs)	*Number*	*Minimum*	*Maximum*	*Mean*	*Standard Deviation*
Total sample	42	1 year 2 months	13 years	3 years 5 months	± 2 years 6 months
A)Younger than 2 years	13	1 year 2 months	1 year 6 months	1 year 4 months	± 2 months
B) 2-4 years	16	2 years	4 years	2 years 7 months	± 8 months
C) Older than 4 years	13	5 years	13 years	6 years 6 months	± 2 years 4 months

Table 3.17: Group A (younger than 2 years): Right ventricular outflow tract obstruction and other cardiac abnormalities occurring in tetralogy of Fallot

No	Age	Sex	Race	Infundibular stenosis	PV stenosis	PDA	ASD
1.	7 months	F	B	Tight	Tight	×	×
2.	1 year 2 months	F	B	√	√	×	×
3.	1 year 2 months	M	B	√	√	×	×
4.	1 year 2 months	F	B	Tight	√	×	4mm
5.	1 year 3 months	F	B	Tight	√	×	×
6.	1 year 4 months	F	I	√	√	2mm	×
7.	1 year 5 months	F	B	√	×	×	×
8.	1 year 5 months	M	W	No comment recorded	√	×	×
9.	1 year 5 months	F	B	Mild	×	×	×
10.	1 year 5 months	F	W	No comment recorded	√	×	×
11.	1 year 6 months	M	C	√	√	√	×
12.	1 year 6 months	M	B	Tight	×	×	×
13.	1 year 6 months	F	B	No comment recorded	√	×	5.8mm

PV=pulmonary valve; PDA=patent ductus arteriosus; ASD=atrial septal defect;
× = absent; √ = present

Table 3.18: Group B (2-4 years): Right ventricular outflow tract obstruction and other cardiac abnormalities occurring in tetralogy of Fallot

No	Age	Sex	Race	Infundibular stenosis	PV stenosis	PDA	Other
1.	2	M	B	√	×	×	×
2.	2	F	C	Moderate	×	×	Discrete subaortic stenosis
3.	2	F	B	No comment recorded	√	×	×
4.	2	F	C	√	√	×	×
5.	2	M	B	Tight	×	×	×
6.	2	M	B	Tight	×	×	×
7.	2	M	B	√	√	×	×
8.	2	F	B	√	√	×	×
9.	3	F	B	No comment recorded	√	×	×
10.	3	M	B	√	√	2mm	
11.	3	M	B	Tight	×	×	×
12.	3	M	B	Tight	×	×	×
13.	3	M	B	√	√	2mm	6mm atrial septal defect
14.	4	M	B	No comment recorded	√	×	×
15.	4	M	W	Severe	×	×	Double outlet right ventricle
16.	4	F	B	Tight	×	×	×

PV=pulmonary valve; PDA=patent ductus arteriosus

× = absent; √ = present

Table 3.19: Group C (older than 4 years): Right ventricular outflow tract obstruction and other cardiac abnormalities occurring in tetralogy of Fallot

No	Age	Sex	Race	Infundibular stenosis	PV stenosis	PDA	ASD
1.	5	F	B	√	√	2mm	7mm
2.	5	F	B	No comment recorded	√	×	×
3.	5	F	C	√	×	×	1mm
4.	5	M	B	√	×	×	×
5.	5	M	C	√	×	×	×
6.	5	M	B	No comment recorded	√	×	×
7.	6	M	B	√	×	×	×
8.	6	M	B	No comment recorded	√	×	×
9.	7	M	B	Tight	×	×	×
10.	9	F	B	Tight	×	×	×
11.	9	F	B	√	√	×	×
12.	13	F	B	√	√	×	×

PV=pulmonary valve; PDA=patent ductus arteriosus; ASD=atrial septal defect;
× = absent; √ = present

Table 3.20: Group A (younger than 2 years): First clinical presentation of the patients with tetralogy of Fallot (TOF) (n=13)

Reason for initial TOF diagnoses	*Number of cases*
Cardiac murmur	5
Respiratory distress + cyanotic spells	5
TOF already diagnosed	3

Table 3.21: Group B (2-4 years): First clinical presentation of the patients with tetralogy of Fallot (n=16)

Reason for initial TOF diagnoses	*Number of cases*
TOF already diagnosed	7
Respiratory distress+ cyanotic spells	5
Cardiac murmur	3
Bronchopneumonia	1

Table 3.22: Group C (older than 4 years): First clinical presentation of the patients with tetralogy of Fallot (n=13)

Reason for initial TOF diagnoses	*Number of cases*
TOF already diagnosed	10
Respiratory distress	2
Cardiac murmur	1

Table 3.23: Type and size of the ventricular septal defect occurring in the cases diagnosed with tetralogy of Fallot

Type	**Group**	**Sample Size**	**No. of measurements**	**Measurement**	**M1**	**M2**	**M3**	**M4**
LARGE PM VSD	Group A	10	4	Echocardiogram	6.6mm	9.9mm	10mm	7mm
	Group B	16	4	Echocardiogram	8.3mm	11mm	11mm	8.7mm
	Group C	12	3	Echocardiogram	20mm	12mm	10mm	-
	Total	38	11					
OUTLET VSD	Group A	3	3	Echocardiogram	7mm	8mm	8.3mm	-
	Group C	1	1	Echocardiogram	18mm	-	-	-
	Total	4	4					

M1, 2, 3, 4= measurement 1, 2, 3, 4

Table 3.24: Age distribution and grouping of the cases with double chambered right ventricle

Age (yrs)	*Number*	*Minimum*	*Maximum*	*Mean*	*Standard Deviation*
Total sample	8	6 months	6 years	2 years 7 months	±1 year 7 months
A) Younger than 2 years	2	6 months	8 months	-	
B) 2-4 years	5	2 years	4 years	2 years 8 months	± 8 months
C) Older than 4 years	1	6 years		-	

Table 3.25: Type and average size of the ventricular septal defect in double chambered right ventricle as documented by the cardiologist from the echocardiogram

	Perimembranous ventricular septal defect n=6		**Outlet n=2**
	Moderate **n=3**	*Small* **n=3**	
Echocardiogram	**7.3mm**	**3.8mm**	**11mm**

4. DISCUSSION

Although the literature is replete with studies describing ventricular septal defects, very little has been documented about these defects in the different population groups in South Africa. This descriptive, retrospective, clinical and anatomical study sheds light on the incidence, anatomical location and size of the different types of ventricular septal defects in a South African group.

In the cadaveric study, all 11 neonates were of the Black South African population group and ranged in age from 24 to 40 weeks. In this group the perimembranous septum and the muscular septum constituted the largest component of the interventricular septum, while the inlet septum and outlet septum contributed the smaller components of the interventricular septum.

In the seven postmortem heart specimens studied, all were from White patients and each demonstrated a perimembranous type of ventricular septal defect. Unfortunately, in most of these cases no clinical information was available, highlighting the need for accurate record keeping and/or the setting up of a subject/patient database in our Faculty. Other abnormalities were noted in association with the ventricular septal defect. In one of the postmortem specimens, in this study, the infant had Down's syndrome. Down's syndrome or Trisomy 21 is a common genetic disorder which often presents with birth defects involving the heart of which a ventricular septal defect is the most common (Krabill, 2004). In another specimen, blood cysts were observed on the tricuspid valve. Blood cysts are diverticuli lined by endothelium and filled with blood (Agac *et al*., 2009). They are usually congenital in origin, and are seen predominantly in infants. Blood cysts are formed by the invagination of crevices on the valve surface into the stroma as a result of high ventricular pressures (Agac *et al*., 2009).

Non-compaction of the left ventricle was seen in one specimen with a ventricular septal defect. The prominent trabeculations in the left ventricle were best seen in the apical and mid-portion of the ventricle. This specimen also had evidence of a dilated cardiomyopathy. Ozgur *et al*. (2011) evaluated 29 children with non-compaction. Of the 29 patients, 13 (45%) were female and 16 (55%) were male. The mean age at presentation was 4.8 years. In three of their cases (10%), ventricular septal defects were also present in addition to the non-compaction. Hunter (2008) studied 100 patients with ventricular septal defects also at the provincial hospitals in Johannesburg, utilizing echocardiography. Non-compaction of the left ventricle (a ratio of greater than 2:1) was found in 42% of her cases. Other studies have also suggested that the presence of non-compaction may portend a poorer prognosis (Weintraub, 2007).

During the analysis of tetralogy of Fallot, a subgroup (n =8; 16%) was identified which is referred to in the literature as double chambered right ventricle. These cases with double chambered right ventricle present with obstruction within the right ventricle which is situated much lower than the right ventricular outflow tract obstruction associated with tetralogy of Fallot. Anismasahun *et al.* (2011) in a case report described only one case of double chambered right ventricle with a ventricular septal defect in an eight year old female with acyanotic congenital heart disease that presented at a teaching hospital in Nigeria. These authors describe double chambered right ventricle as an uncommon congenital heart disease. However, in the present study, there was a concurrent presentation of double chambered right ventricle with a ventricular septal defect in 16% of cases. The male-to-female ratio in double chambered right ventricle is 2:1 (Rowland *et al*., 1975; Telagh *et al*., 2008; Anismasahun *et al.,* 2011). Similarly

in the small sample of double chambered right ventricle in the present study, there were also more males than females (3:1).

The most common reason for referral of a double chambered right ventricle is the detection of a murmur (Anismasahun *et al.,* 2011). A cardiac murmur was heard at birth in five of the eight cases (63%) with double chambered right ventricle in this series. Patients with double chambered right ventricle usually present with left to right shunting of blood. Seventy five percent (six out of eight) of patients in this study had left to right shunting of blood, compared to an American study by Rowland *et al*. (1975) in which 41% of patients had left to right shunting of blood.

The most common cardiac defect that occurs in association with double chambered right ventricle is the perimembranous ventricular septal defect (Anismasahun *et al.,* 2011). In the present study, in the Black South African population, 75% of the eight cases with double chambered right ventricle had a perimembranous ventricular septal defect while Telagh *et al*. (2008) in a German population observed ventricular septal defects in 70% of their patients with double chambered right ventricle. Rowland *et al*. (1975), in a study conducted on 17 American patients, reported an even higher incidence of ventricular septal defects with double chambered right ventricle (88% of the 17 cases).

Subaortic stenosis and pulmonary valve stenosis may also be associated with double chambered right ventricle (Rowland *et al*., 1975). Discrete subaortic stenosis was found in 38% (three out of eight) of cases with double chambered right ventricle in the present study and in only 12% (eight out of 17) of patients in the study by Rowland *et al.* (1975). In contrast, in an echocardiographic

study by Vogel *et al.* (1988), of the 36 patients with the combination of double chambered right ventricle and a perimembranous ventricular septal defect, there was an associated subaortic abnormality in 32 (88%) cases. Thus, these authors concluded that the incidence of discrete subaortic stenosis associated with double chambered right ventricle appears to be far greater than previously suspected (Vogel *et al.*, 1988). Most authors in the literature (Rowland *et al*., 1975; Telagh *et al*., 2008; Anismasahun *et al.*, 2011) conclude that due to the developmental and progressive nature of double chambered right ventricle, early surgical intervention to correct the obstruction and to close the ventricular septal defect is warranted.

The predominant population group in all three of the subgroups in the present study was Black. This study was carried out on surgical closures of ventricular septal defects between 2001 and 2004 but demographic changes have occurred in South Africa since 1994 resulting in a predominance of Black children undergoing surgery at the provincial hospitals in Johannesburg. The study thus does not necessarily indicate a higher prevalence of ventricular septal defects in the Black South African population. This is in contrast to the results of a study in 1990 by Klass *et al*. where almost equal numbers of white and black children were operated upon.

With regards to sex incidence, there were more females than males in the cases with isolated ventricular septal defects, equal numbers of males and females in the cases with the tetralogy of Fallot, and, in contrast to the cases with double chambered right ventricle where more males were represented than females. The present study accords with Hoffman and Rudolph (1965), Klass *et al*. (1990) and Krabill (2004) who all reported a higher incidence of ventricular septal defects in females. The present study concurs with that of Shinebourne and Anderson (2002)

who reported that males and females are equally affected in the tetralogy of Fallot, and also with the review by Anismasahun *et al.* (2011) who found that the incidence of ventricular septal defects in double chambered right ventricle was more common in males. In the present study, a predominance of males was also found.

In relation to the first presentation of the isolated ventricular septal defect, the results of the present study show that the ventricular septal defect was already diagnosed at a pre-hospital visit in eleven of the 50 cases (22%), while a higher percentage, 48% (20 out of 42 cases) were referred to the cardiac clinic with a diagnosis of tetralogy of Fallot. In contrast, only one of the eight cases (13%) with double chambered right ventricle was recognised prior to attendance at the cardiac clinic. With the demographic changes that have occurred in South Africa post-apartheid, the overall care of patients has also changed. Thus most cases for surgical correction are placed on a waiting list for an operation date (91% of cases in the present study). In the Klass *et al*. (1990) study, 71% of cases underwent surgery under the age of two years compared to the present study where only 40% of cases under the age of two years underwent surgery.

In the present study, 90% of cases had a single ventricular septal defect while in the remaining 10% there were multiple ventricular septal defects. In all the cases analyzed, a perimembranous ventricular septal defect was the most common type diagnosed; 78% in the cases with a single isolated ventricular septal defect, 90% in patients with tetralogy of Fallot and 75% in the group with double chambered right ventricle. A perimembranous ventricular septal defect was also the most common type diagnosed in the cases where more than one type of defect occurred.

The results of this study compare favorably with those of other studies on surgery for ventricular septal defects. In the previous South African study by Klass *et al*. (1990) a perimembranous ventricular septal defect was also the most common, with identical percentages (65%) for White and Black children. In a British study, on 220 patients by Soto *et al*. (1980), perimembranous ventricular septal defects were found in 70% of cases, while Santamaria *et al*. (1983) reported an incidence of 82% in another British study. Anderson *et al*. (1981) reported an incidence of 79% of perimembranous ventricular septal defects associated with tetralogy of Fallot. This is lower than the 90% found in the present study for the cases with tetralogy of Fallot, but this may be accounted for by sample size and selection of patients.

In the cases with isolated ventricular septal defects, in the present study six infants had a muscular ventricular septal defect (13%). This result is higher than that of Klass *et al.* (1990) (5%) and the 8% reported by Santamaria *et al*. (1980). In the present study, three infants in the cases with isolated ventricular septal defects were diagnosed with outlet ventricular septal defects (7%) and only one child had an inlet ventricular septal defect (2%). In the cases with tetralogy of Fallot, 10% had an outlet type of ventricular septal defect. This result contrasts with the results in the Anderson *et al*. (1981) study, where outlet ventricular septal defects were reported in 21% of 53 cases. In the cases with double chambered right ventricle, 25% had an outlet type of ventricular septal defect.

No sub-arterial defects were recorded in this study as well as in the study by Klass *et al*. (1990). These defects could have possibly been recorded as an outlet type of ventricular septal defect. This contrasts with the situation in Japan where there is a high prevalence of sub-arterial

ventricular septal defects (10% to 20%) (Tatsuno *et al*., 1973a; Soto *et al*., 1980; Ando *et al*., 2003).

During operation, the anatomical location and size of the ventricular septal defects were recorded by the surgeon in the operative reports. The perimembranous ventricular septal defects were described as large, moderate and small in size by the surgeon in the operative reports. It is important to note that the size of these defects was an "eyeball" estimation made by the surgeon and was not directly measured, hence these measurements were not used for analysis. It is also important to note that during the analysis of the echocardiographic and operative reports there was missing data with regard to the record of the size of the ventricular septal defect. The need for proper record keeping is once again highlighted.

A large perimembranous ventricular septal defect was found in twenty of the cases with isolated ventricular septal defects (11.7 mm from echocardiogram) and also in the 38 cases with tetralogy of Fallot (10.4 mm from echocardiogram). Large isolated ventricular septal defects can cause heart failure and poor growth in infants. Surgery to close the defect is mandatory (Krabill, 2004).

The moderate and small perimembranous ventricular septal defects were seen in the cases with an isolated ventricular septal defect. Seven were moderate (7.6 mm from echocardiogram), and eight were small (5.8 mm from echocardiogram). In the cases with double chambered right ventricle three were moderate (7.3 mm from echocardiogram), and three were small (3.8 mm from echocardiogram). Small ventricular septal defects can undergo spontaneous closure, but periodic follow up is necessary where spontaneous closure does not occur (Krabill, 2004).

Patients with small defects have an excellent long term prognosis with no risk of developing pulmonary vascular disease. The patients with moderate sized defects do not have heart failure, but elective surgery is necessary to prevent the development of pulmonary vascular disease (Krabill, 2004).

The average size of the outlet type of ventricular septal defect was large and was similar for all three of these patients (between 10 mm and 11 mm). The six muscular type of ventricular septal defect was found only among the isolated ventricular septal defect and measured 9.3 mm from the echocardiogram. The one inlet ventricular septal defect measured 9 mm from the echocardiogram.

The technical advances in echocardiography have allowed for accurate measurements in the size of ventricular septal defects in patients.

In the five cases with multiple ventricular septal defects, all the measurements for the perimembranous ventricular septal defect were recorded, but the size of the associated smaller muscular ventricular septal defects was not recorded.

The haemodynamic consequence of the ventricular septal defect was analysed from the reports. In the cases with an isolated ventricular septal defect and in double chambered right ventricle, left to right shunting was the predominant haemodynamic consequence of the ventricular septal defect (82% and 75% respectively). Normally the systolic left ventricular pressure is about four times greater than the systolic right ventricular pressure (Spencer, 1984). In tetralogy of Fallot

right to left shunting and bi-directional shunting was the predominant finding (40% and 36% respectively) due to the obstruction of the right ventricular outflow tract together with the ventricular septal defect.

A large ventricular septal defect results in a left to right shunt of blood from the left ventricle into the pulmonary circulation producing a pulmonary blood flow at least twice or more than the systemic blood flow. In the small ventricular septal defects the pulmonary blood flow is only about one and a half times the systemic blood flow and few physiological changes result (Spencer, 1984). These small defects often undergo spontaneous closure and are not of great concern to the surgeon. If a moderate to large ventricular septal defect is not operated on in the first two years of life, the pulmonary hypertension eventually damages the pulmonary vasculature so that pulmonary vascular resistance exceeds the systemic vascular resistance, and a right to left shunt occurs. This advanced inoperable condition with irreversible pulmonary vascular disease is termed the Eisenmenger syndrome (Spencer, 1984; Krabill, 2004).

From the cardiac catheterization data (see Table on Page 148 in Appendix A) a large left to right shunt ,where the Qp:Qs ratio is equal to or greater than 2:1 was found in 63% of patients. Further confirmation of a significant VSD was obtained from the pulmonary artery systolic pressure which was 40 mm Hg or greater in 77% of the subjects. Of interest, a Qp:Qs ratio of 0.9 was recorded in three patients, two of whom had very high pulmonary systolic pressures. This observation may be an indirect indication of the beginning of a right to left shunt (Eisenmenger Syndrome). The other patient with a Qp:Qs of 0.9 had a surprise finding of a low (20mm Hg) pulmonary artery systolic pressure. This observation is unexplained.

Additional cardiac abnormalities that occurred in association with the ventricular septal defect were recorded and analyzed as part of this study. Prolapse of the right coronary cusp of the aortic valve into the ventricular septal defect was documented in 22% of patients with an isolated ventricular septal defect and aortic regurgitation occurred in 54.5% of these cases. Nadas *et al.* (1964) conducted a study in an American population on 756 patients with ventricular septal defects. Thirty four (4.6%) of them were diagnosed with associated aortic regurgitation. In comparison, a study by Tatsuno *et al.* (1973a) in a Japanese population, documented ventricular septal defects associated with aortic regurgitation in 8.2% of cases. Furthermore, Tatsuno *et al.* (1973a) found that in Japan the position of the VSD is supracristal (subarterial) in most cases (78% of their 91 cases). However, according to American (Van Praagh and McNamara, 1968) and British reports (Somerville et al., 1970), the number of supracristal ventricular septal defects with aortic regurgitation is approximately the same as that of infracristal ventricular septal defects with aortic regurgitation. Tatsuno *et al.* (1973a) propose that one of the reasons for such a difference is that in Japan, the number of uncomplicated supracristal ventricular septal defects is greater than the number reported in other countries. However, the intrinsic cause is due to a difference in the development of the conus.

There is only one recent study describing this anomaly in South Africa. Pepeta *et al.* (2006) reviewed 101 patients who underwent surgery for a ventricular septal defect between 2000 and 2005. Of these cases, 20 (19.8%) were found to have prolapse of the aortic leaflet (12 of the 20 had prolapse of the right coronary cusp while four were from the non-coronary cusp). This incidence of 19.8% compares favorably with the 22% of cases with aortic cusp prolapse found in the present study. In the Pepeta *et al*. (2006) study, all 20 of the patients with prolapse had aortic

regurgitation (trivial or mild regurgitation in 14 cases). Of the six with moderate to severe regurgitation, three underwent aortic valve surgery.

Strangely, there was no documentation of prolapse among the Black children in the previous report by Klass *et al*. (1990). However the study by Klass *et al.* (1990) was a clinical study with no echocardiographic input. Hence, aortic regurgitation may not have been diagnosed. Based on the results of the present study and those of Pepeta *et al*. (2006), it appears that there is a higher incidence of prolapse of the aortic cusp (19.8%-22%) and aortic regurgitation in the Black patients than that reported in Western countries (4.6%-9.8%) (Nadas *et al.*, 1964; Van Praagh and McNamara, 1968; Somerville *et al*., 1970), or in Asia (8.2%) (Tatsuno *et al*., 1973a).

In the present study, atrial septal defects occurred in association with a ventricular septal defect in 11% of cases (six in the group with an isolated ventricular septal defect; five in tetralogy of Fallot). In comparison, atrial septal defects occurred in association with tetralogy of Fallot in 38.4% of 158 cases in a British study conducted by Suzuki *et al*. (1990).

The anomaly of double outlet right ventricle is a cardiac malformation in which both the aorta and the pulmonary artery arise from the right ventricle (Cavalini *et al*., 1998). Two of the three cases with double outlet right ventricle in this study had isolated ventricular septal defects and one case had tetralogy of Fallot. Similarly Cavalini *et al*. (1998) in their study described two cases with this rare phenomenon.

Discrete subaortic stenosis was noted in fifteen patients, eleven of the cases had an isolated ventricular septal defect, three occurred in association with double chambered right ventricle and

the remaining one case had tetralogy of Fallot. In the cases with tetralogy of Fallot, the right ventricular outflow tract obstruction was subvalvular (infundibular) in 76% of patients in the present study compared to the 44% reported by Vargas *et al*. (1986) in a study conducted in Argentina. Pulmonary valve stenosis occurred in 57% of cases in the present study which is similar to the 51% of patients reported by Hirsch *et al.* (2000) in an American study. In tetralogy of Fallot, the antero-cephalad displacement of the infundibular septum results in the underdevelopment of the infundibulum with pulmonary outflow tract obstruction (Van Praagh *et al*., 1970). If infundibular underdevelopment is moderate, stenosis of the pulmonary outflow tract is moderate and if infundibular underdevelopment is marked, extreme stenosis of the pulmonary outflow tract results (Van Praagh *et al*., 1970). Early relief of the right ventricular outflow tract obstruction may result in early resolution of the right ventricular hypertrophy and fibrosis and subsequently decreases the incidence of right ventricular dysfunction (Hirsch *et al.,* 2000).

Early repair of a ventricular septal defect has many advantages. Especially in the cases with tetralogy of Fallot, early repair abolishes the secondary effects of cyanosis on vital organs and the adverse effect that it has on cognitive and psychomotor development (Caspi *et al*., 1999). In the isolated ventricular septal defects, early repair is required before the onset of Eisenmenger's syndrome. When investigating a ventricular septal defect it is simply not enough to diagnose the presence of a "hole" in the interventricular septum. A thorough investigation including careful echocardiography should identify the defect i.e. defining its site, size and the structures forming its margins. This will enable an accurate morphological classification and will facilitate the appropriate and necessary surgical approach.

5. CONCLUSION

The primary purpose of this study was to retrospectively investigate two groups of classified patients who had undergone surgery for the closure of (i) isolated ventricular septal defects (50 patients) and (ii) repair of tetralogy of Fallot (50 patients). Particular attention was given to the ventricular septal defects recorded in these cases. This review of surgery for isolated ventricular septal defects is only the second such study conducted in this cardiovascular unit.

The developmental process and the complex nature of the formation of the pars membranacea of the interventricular septum explains why the most common ventricular septal defect found in this study is that of a perimembranous type. The detailed study of the normal neonatal interventricular septum highlighted the morphology of the region. The results show that the various components of the interventricular septum are larger in neonates of a more advanced gestational age.

Of the cases that presented for operation, 90% in the group with isolated ventricular septal defects were single defects. Anatomically the most common site was perimembranous (78%). The other sites were classified as muscular or occurred in the outlet region of the right ventricle.

An important additional finding of this study was prolapse of the right coronary cusp of the aortic valve into the ventricular septal defect (22% of the patients) and consequential aortic

regurgitation in half of these patients. This prevalence is greater than that reported in the literature for White and Japanese children.

Analysis of the patients classified as tetralogy of Fallot revealed an important subgroup (eight patients) referred to as double chambered right ventricle. In contrast to those with tetralogy of Fallot, the ventricular septal defect was variable in size and not as large as that found in those infants with tetralogy. Furthermore the site of right ventricular obstruction is due to muscle bundles lower than in the right ventricular outflow tract.

Non-compaction of the left ventricle was seen in one post-mortem specimen in association with a perimembranous ventricular septal defect and dilated cardiomyopathy. The presence of this condition in association with a ventricular septal defect has been reported to worsen the prognosis for survival.

Appendix A: **Listing of individual results**

Table 1: Age distribution of the cases that presented with a VSD as an isolated defect

Age (yrs)	***Number***	***%***	***Minimum***	***Maximum***	***Mean***	***Standard Deviation***
Total sample	*50*	*100*	*0.42*	*13*	*3.14*	*2.97*
< 1 year	9	18	0.42	0.83	0.61	0.14
1-4 years	28	56	1	4	2.01	0.96
> 4 years	13	26	5	13	12.12	2.68

Table 2a:

Weight (kg)	***Total sample n=50***	**< 1 year n=9**	**1-4 years n=28**	**> 4 years n=13**
Minimum	*3.3*	3.3	4.7	13.5
Maximum	*31*	6.2	14.8	31
Mean	*10.54*	4.77	9.39	17.02
Standard deviation	*5.35*	1.10	2.63	5.20

Table 2b:

Height (m)	***Total sample n=50***	**< 1 year n=9**	**1-4 years n=28**	**> 4 years n=13**
Minimum	*0.53*	0.53	0.60	0.84
Maximum	*1.34*	0.71	1.03	1.34
Mean	*0.80*	0.62	0.80	1.12
Standard deviation	*0.29*	0.22	0.19	0.35

Prenatal history

Normal vs. complicated birth

Prenatal history	
Normal births n= 47(94%)	*Complicated births n= 3(6%)*

• Infants were born at term by normal vaginal delivery. • No complications.	Premature birth (n=1) • 6 month old black female • One of twins • Systolic murmur heard at birth	Births by caesarian section (n=2) 1) Due to poor prognosis • 8 month old black female • breathing difficulties at birth 2) Due to foetal distress • 2 year old black female

Previous operation details

Previous operation details n= 4 (8%)					
No.	*Age (yrs)*	*Sex*	*Race*	*Operation details*	*Complications*
1.	1	M	Black	MPA banding	Failed extubation 5 times
2.	1	M	White	a) Closure of PM VSD (12-13mm) using a vascular goretex patch. b) PDA ligation. c) Excision of atrial septum with rerouting of the left SVC into the RA using a bovine pericardial patch.	• High PA & LA pressure • Coronary sinus patch protruded into LA. • PHT due to residual VSD • Pulmonary venous obstruction caused by ASD patch.
3.	1	F	Black	PDA ligation	No complications
4.	4	M	Black	a) Repair of a coarctation of the aorta with a subclavian flap. b) MPA banding	•Dense adhesions formed in the region of the band. • The band was very tight and was impinging on the RPA.

Diagnosis associated with the VSD

Diagnosis of the type of VSD in cases where only 1 VSD was present

Table 3: Types of VSD in cases diagnosed with only 1 type of VSD

Types of VSDs (cases where single VSDs occurred) n=45 (90%)								
	PM VSD		Muscular VSD		Outlet VSD		Inlet VSD	
	No.	%	No.	%	No.	%	No.	%
Total sample (n=45)	*35*	*77.8*	*6*	*13.3*	*3*	*6.7*	*1*	*2.2*
<1 year (n=8)	6	75	1	12.5	1	12.5	-	-
1-4 years (n=25)	20	80	3	12	1	4	1	4
> 4 years (n=12)	9	75	2	16.7	1	8.3	-	-

Diagnosis of the type of VSD in cases where multiple VSDs were present

No.	Age	Gender	Race	VSDs
1.	7 months	Female	White	PM & apical muscular
2.	1 year	Female	Black	PM & apical muscular
3.	1 year	Female	Black	PM, mid muscular & Swiss cheese
4.	1.5 years	Female	Black	PM & mid muscular
5.	12 years	Female	Black	PM & 2 mid muscular

Physiological diagnosis of the VSDs in cases where only 1 VSD was present

Table 4: Physiological diagnosis of the VSDs where only 1 type of VSD occurred

Perimembranous VSD- physiological diagnosis						
	Right left shunting		Left to right shunting		Bi-directional shunting	
	No.	%	No.	%	No.	%
Total sample (n=35)	-	-	*30*	*85.7*	*4*	*8.6*
< 1 year (n=6)	-	-	3	50	2	33.3
1-4 years (n= 20)	-	-	19	95	1	5
> 4 years (n= 9)	-	-	8	88.9	1	11

Muscular VSD- physiological diagnosis						
	Right to left shunting		Left to right shunting		Bi-directional shunting	
	No.	%	No.	%	No.	%
Total sample (n=6)	-	-	4	66.7	2	16.7
< 1 year (n=1)	-	-	1	100	-	-
1-4 years (n=3)	-	-	2	66.7	1	33.3
> 4 years (n=2)	-	-	1	50	1	50

Outlet VSD- physiological diagnosis						
	Right to left shunting		Left to right shunting		Bi-directional shunting	
	No.	%	No.	%	No.	%
Total sample (n=3)	-	-	3	100	-	-
< 1 year (n=1)	-	-	1	100	-	-
1-4 years (n=1)	-	-	1	100	-	-

Inlet VSD- physiological diagnosis						
	Right to left shunting		Left to right shunting		Bi-directional shunting	
	No.	%	No.	%	No.	%
Total sample (n=1)	-	-	-	-	-	-
< 1 year (n=0)	-	-	-	-	-	-
1-4 years (n=1)	-	-	-	-	-	-
> 4 years (n=0)	-	-	-	-	-	-

Physiological diagnosis of the VSDs in cases where multiple VSDs were present

No.	**Age**	**Gender**	**Race**	Physiological diagnosis
1.	7 months	Female	White	•Step up of O sats by 40% from SVC to RA suggests a left to right shunt consistent with ASD. •Increased arterial O sats in MPA consistent with a left to right shunt through VSD/PDA.
2.	1 year	Female	Black	•Step up in O sats from RA 71% to RV 81% indicative of L to R shunting at ventricular level.
3.	1 year	Female	Black	• Bi- directional shunting of blood
4.	1.5 years	Female	Black	•Step up in sats from RA to RV & RV to MPA suggestive of VSD & PDA & L to R shunting
5.	12 years	Female	Black	•Step up in O sats in RA & RV due to L to R shunting. No significant drop in LV sats from LA due to R to L shunting across VSD i.e. bi-directional

Other cardiac abnormalities

Table 5: Other cardiac abnormalities: < 1-year age group

Cases with only type of VSD						
No	Age	Sex	Race	General	Atrium	Ventricle
1.	0.42	M	Black	Cardiomegaly with apex beat in the 5th i.c.s. All 4 chambers of heart dilated. 2-3mm PDA. PFO.	Interatrial septum thickened.	
2.	0.5	F	Black	All 4 chambers of heart dilated, apex displaced to 6th l.i.c.s.		
3.	0.5	M	Black	No PDA only 1mm ligamentum seen, 4-5mm PFO, all 4 chambers of heart dilated, cardiomegaly with apex beat in 5th i.c.s.	4mm ASD secundum	Biventricular enlargement
4.	0.5	M	Indian	3mm PDA, all 4 chambers of heart dilated.		Biventricular hypertrophy.

5.	0.58	F	White	Cardiomegaly with apex beat in 5th i.c.s. Situs solitus. 2mm PDA.	Secundum ASD situated superiorly under left atrial appendage & was 6mm (opp) & 9mm (echo). Inferiorly large coronary sinus draining into RA. Cor triatriatum (suspicion of membrane in LA proximal to ASD), atrial appendages juxtaposed on right with left atrial appendage situated superiorly & right atrial appendage situated inferiorly, RA dilated. LA small.	RV dilated with strange configuration, looked like apex pointing towards the right.
6.	0.67	F	Black	All 4 chambers of heart significantly enlarged. Cardiomegaly with apex displaced to 6th l.i.c.s.		
7.	0.75	F	Black	Situs solitus. Mild cardiomegaly with apex beat in 5th i.c.s. & slightly displaced.	5mm fenestrated secundum ASD.	
8.	0.75	F	Black	Situs solitus. PFO= 2-3mm. Cardiomegaly with apex on 5th i.c.s. All 4 chambers of heart dilated. Mid diastolic murmur at apex. Mild mitral regurgitation.	-	Biventricular enlargement.
9.	0.83	F	Black	Situs solitus. Prolapse of aortic cusp in VSD. All 4 chambers of heart dilated. Cardiomegaly with apex beat in 5th i.c.s. lateral to MCL.	-	Bi-ventricular hypertrophy.

Table 6& 7: Other cardiac abnormalities: 1-4 years age group n=28 (56%)

Other cardiac abnormalities						
No.	Age	Sex	Race	General	Atrium	Ventricle
1.	1	M	White	Situs solitus, Cardiomegaly with the apex beat in the 5th i.c.s.	Secundum ASD=7.8mm (echo), 8mm (op), RV hypertrophied	-
2.	1	F	Black	The 4 chambers of the heart were dilated.	-	-
3.	1	M	Black	Situs solitus. Cardiomegaly with the apex beat in the 6th i.c.s. There were loose, hemorrhagic adhesions between heart & pericardium. The 4 chambers of the heart were dilated. The MPA band did not appear very tight. No mid diastolic murmur. There was a tiny 1 mm PFO	-	RV hypertrophied
4.	1	M	White	Situs solitus. PDA=3mm (cath), 4-5mm (op). Heart was markedly dilated rotated towards the right (mesocardia). Marked 4 chamber enlargements.	ASD present. Left SVC draining into roof of LA	RV heavily trabeculated.
5.	1	F	Black	The 4 chambers of the heart were dilated. No mid diastolic murmur. Apex in the 5th i.c.s. Lateral to MCL. PDA=4mm	-	-

6.	1	F	Black	4 chambers of the heart were significantly dilated. . Apex in the 6th I.c.s. outside MCL. Small PFO = 2mm	LA dilated	Biventricular enlargement
7.	1	M	Black	Situs solitus, Cardiomegaly with apex beat in the 5th i.c.s lateral MCL. Abnormal systemic venous return	Enlarged RA.	Bi-ventricular enlargement. RV not well trabeculated but of adequate size.
8.	1	M	Indian	Situs solitus. 4 chambers of the heart were dilated. No mid diastolic murmur. Cardiomegaly with the apex beat in the 5th i.c.s. PDA=4mm. Tiny PFO. Signs of CCF	-	-
9.	1.2	F	Black	Situs solitus. Mild cardiomegaly	-	-
10.	1.5	F	Black	Large PDA. Apex beat displaced into the 6th I.c.s. lateral to mid clavicular line.	-	-
11.	1.67	M	Black	Situs solitus. Cardiomegaly with the apex displaced onto the 6th i.c.s, mild MR due to prolapse of the AMVL	-	Biventricular enlargement
12.	2	F	Black	Situs solitus, discrete subaortic stenosis posterior malaligned aorta with subaortic tunnel, 4 chambers dilated, Cardiomegaly, apex in 5th ICS	Left atrial enlargement	Biventricular hypertrophy
13.	2	F	Black	4 chambers of the heart were dilated. Cardiomegaly with apex beat in the 6th LICS lateral to MCL.	Large LA with the inter atrial septum bulging to the right	Biventricular hypertrophy
14.	2	F	Black	Cardiomegaly with the apex beat in the 6th i.c.s. Minor aneurysm of right sinus of valsalva- not ruptured	Dilated LA. Enlarged RA. Interatrial septum intact	Dilated LV
15.	2	F	Black	Moderate valvar PS Moderate to severe tricuspid regurgitation	Large secundum ASD=19mm,practically a common atrium with only the medial rim still present.	-
16.	2	M	Black	Prolapsed RCC into the VSD. Cardiomegaly with the apex beat in the 5th i.c.s. Signs of CCF.	RA dilated & enlarged. Interatrial septum intact	Biventricular enlargement
17.	2	M	Black	PDA ligamentum=3mm. 4 chambers of the heart significantly dilated.	-	-
18.	2	M	Black	All 4 chambers of the heart were dilated. Cardiomegaly, apex is localised in the 6th left i.c.s. lateral to MCL.	-	Biventricular hypertrophy
19.	2	M	Black	Cardiomegaly with the apex beat in the 5th i.c.s. All 4 chambers of the heart were dilated. Discrete subaortic stenosis. Moderate aortic regurgitation. Prolapse of the RCC into the VSD	-	-
20.	2	F	Black	The 4 chambers of the heart were dilated. LPA stenosis	-	
21.	2	M	Black	DSAS, Cardiomegaly with apex beat in the 5th i.c.s. lateral MCL.	-	Bi-ventricular hypertrophy
22.	3	F	Black	Small PFO = 2mm. PDA was small measuring approximately 2mm. Cardiomegaly with apex in the 6th l.i.c.s. Significant 4 chamber enlargement.	-	LV dysfunction, Contractility is not normal

23.	3	F	Indian	Cardiomegaly with apex in the 6th l.i.c.s. Systolic thrill	Interatrial septum intact but bulges to the right	Bi ventricular hypertrophy
24.	3	M	Black	The 4 chambers of the heart were dilated, Small PDA 3-4mm., Apex displaced to 6th l.i.c.s. Discrete subaortic stenosis	LA dilated	Biventricular hypertrophy
25.	3	F	Black	Cardiomegaly with the apex beat in the 5th i.c.s. The 4 chambers of the heart were dilated.	Dilated L & R atria	-
26.	4	F	Black	PDA= 3-4mm. The 4 chambers of the heart were dilated. Cardiomegaly with apex beat displaced to 6th l.i.c.s. lateral to MCL.	-	Bi-ventricular hypertrophy. Double outlet LV. RV mildly hypoplastic
27.	4	M	Black	Apex beat is displaced, Bicuspid aortic valve, Subaortic narrowing due to PM septum, LVOT obstruction, Mild AR	-	-
28.	4	F	Black	Moderate aortic regurgitation due to prolapse of RCC into VSD, DSAS=3.3mm. 4 chambers only mildly dilated.	-	-

Table 8: Other cardiac abnormalities: > 4 years age group n=13 (26%)

Other cardiac abnormalities						
No.	Age	Sex	Race	General	Atrium	Ventricles
1.	5	M	Black	Cardiomegaly with the apex beat in the 6th I.c.s. lateral to the MCL. Enlargement of all 4 chambers. Discrete subaortic stenosis DCRV. Prolapse of aortic cusp into the VSD	-	There was no obvious muscle bundle in RV (op). Echo shows muscle bundle. Biventricular hypertrophy.
2.	5	M	Black	Large amount of old blood was found in the pericardium. All 4 chambers of the heart were markedly dilated.	-	-
3.	5	M	Black	All 4 chambers of the heart were significantly enlarged. PDA=3-4mm. Small PFO.	-	-
4.	5	M	Black	Mild aortic regurgitation due to prolapse of AV leaflet into VSD Discrete sub aortic membrane. Apex beat in the 5th i.c.s. medial to the MCL.	RA enlarged	Biventricular hypertrophy
5.	6	F	Black	PDA=4mm. Signs of CCF with a raised JVP. Cardiomegaly with the apex beat in the 6th i.c.s. The 4 chambers of the heart were significantly dilated	-	Both ventricles are dilated & dysfunctional.
6.	6	F	Black	2 AV valves. Mitral valve cleft with mild to moderate regurgitation & straddling of mitral chordae to IV crest. Cardiomegaly with apex beat in 6th ICS beyond MCL.	ASD secundum =7mm (echo), 12mm by 5mm (op).	-

7.	7	F	Black	Cardiomegaly, apex in 5th l.i.c.s. lateral to MCL. PFO=1mm. Prolapse of RCC into the VSD	-	Bi-ventricular hypertrophy
8.	7	F	Black	4 chambers of the heart were dilated. Cardiomegaly with the apex beat in the 5th l.i.c.s laterally displaced. Discreet subaortic stenosis	-	Biventricular hypertrophy
9.	7	F	Black	Cardiomegaly with apex beat 6th i.c.s. laterally displaced. CCF with elevated JVP. Mid diastolic murmur at apex with pre systolic accentuation. Discrete sub aortic membrane 3.5mm. Aorto-mitral continuity & sub-pulmonary conus. Supra mitral membrane present with central orifice of 4-5mm in diameter & attached to posterior mitral annulus but tethered to para-annular area of AML. MV underneath membrane looked normal. Tight mitral stenosis with mitral opening of 5-7mm due to congenital supra-mitral membrane.	RA + LA dilated.	RV dilated. Double outlet RV.
10.	7	F	Black	The 4 chambers of the heart were mildly enlarged. Prolapsed RCC into the VSD. DSAS. Apex in the 5th i.c.s.	-	-
11.	10	F	Colored	All 4 chambers of the heart were dilated. Cardiomegaly present with apex beat in 6th intercostal space lateral to MCL. DSAS, Partial closure of VSD by aortic valve	-	-
12.	12	F	Black	The 4 chambers of the heart were only mildly enlarged. PFO=5mm	-	-
13.	13	F	Black	4 chambers of the heart were mildly dilated. Cardiomegaly with the apex beat in the 5th l.i.c.s. Aortic regurgitation due to prolapse of the right coronary cusp into the VSD.	-	Left ventricular hypertrophy

Table 9: Genetic abnormalities & syndromes and other abnormalities: < 1 year age group n = 9 (18%)

Other diagnoses: < 1 year age group n=9 (18%)					
No.	Age	Sex	Race	Genetic abnormalities & syndromes	Other abnormalities
1.	0.42	M	Black	-	Dysmorphic features with facial asymmetry. Gastro oesophageal reflux. Epigastric pulsations. Hepatomegaly 2cm. Right LMN 7th cranial nerve palsy
2.	0.5	F	Black	-	Severe failure to thrive. One of premature twins.
3.	0.5	M	Black		Oxygen dependent. Mild oral thrush. 4cm hepatomegaly. No spleen.
4.	0.5	M	Indian	-	Interrupted IVC with azygos continuation. Epigastric heave. Bulbous nose. Prominent ridge of metopic suture suggesting craniostenosis of this suture.
5.	0.58	F	White	-	Very large thymus gland. No innominate veins
6.	0.67	F	Black		Chronic gastro oesophageal reflux. Severe failure to thrive. Dysmorphic features: flat facial profile, hypotelorism, low set ears and a feeble cry. 2 cm liver. No spleen.

7.	0.75	F	Black	-	Syndactyly of small toe & 4th toe on each foot.
8.	0.75	F	Black	-	-
9.	0.83	F	Black	-	2cm palpable liver below the right costal margin.
Cases were more than 1 type of VSD was present					
1.	0.58	F	W	No other abnormality was found	

Table 10 & 11: Genetic abnormalities & syndromes and other abnormalities: 1-4 year age group n=28 (56%)

Other diagnoses: 1-4 year age group n=28 (56%)					
No	Age	Sex	Race	Genetic abnormalities & syndromes	Other abnormalities
1.	1	M	White	-	Failure to thrive Congenital CMV infection, Hepatomegaly 3cm. Splenomegaly 2cm.
2.	1	F	Black	-	-
3.	1	M	Black	-	Hepatomegaly 4-5cm pushed down. Delayed motor milestones due to chronic illness
4.	1	M	White	VACTERL association HOLT-ORAM syndrome	Mild dysmorphism, developmental delay & poor weight gain. Limb anomalies: radial ray abnormalities including a clubbed left hand with a rudimentary thumb & hypoplastic adducted thumb on right hand, radial aplasia on left hand, small thumb on right. Severe equinovarus deformity on right foot. Agenesis of left kidney, abnormal shaped bladder. Congenital ptosis of right eye & craniosynostosis. No innominate vein
5.	1	F	Black	-	-
6.	1	F	Black	-	3cm hepatomegaly. Tip spleen
7.	1	M	Black	-	3cm hepatomegaly. Asymmetrical crying facies
8.	1	M	Indian	-	Epigastric pulsation. Hepatomegaly 4cm
9.	1.2	F	Black	-	-
10.	1.5	F	Black	Down's syndrome	Several large lymph nodes in the axillary area. Very floppy CNS
11.	1.67	M	Black	-	Suspected bilateral SVC's, Dysmorphic features, prominent forehead, deep set eyes, fish mouth. Hypopigmented skin on left upper arm. No spleen.
12.	2	F	Black	-	-
13.	2	F	Black	-	-
14.	2	F	Black	-	-
15.	2	F	Black	-	Bilateral SVC's absent innominate vein
16.	2	M	Black	-	Hepatomegaly 2cm.
17.	2	M	Black	-	-
18.	2	M	Black	-	-
19.	2	M	Black	-	-
20.	2	F	Black		
21.	2	M	Black	-	-
22.	3	F	Black	-	Puffing looking face, metopic suture, mild digital clubbing, Hepatomegaly = 4-5cm. Kidneys, bladder and ureters show a clubbed pelvo-calyceal system on right and asymmetrical bladder. SBE: blood culture positive for streptococcus viridans
23.	3	F	Indian	-	-
24.	3	M	Black	-	-
25.	3	F	Black	-	-
26.	4	F	Black	-	Mild dysmorphic features, bulbous nose with square nasal tip. Bilateral epicanthic folds & relatively small mouth
27.	4	M	Black	-	Mild clubbing of the digits.
28.	4	F	Black	-	-

Table 12: Genetic abnormalities & syndromes and other abnormalities: > 4 years age group n=13 (26%)

Other diagnoses: > 4 years age group n=13 (26%)					
No.	Age	Sex	Race	Genetic abnormalities & syndromes	Other abnormalities
1.	5	M	Black	-	Terrible dental caries
2.	5	M	Black	-	2cm hepatomegaly
3.	5	M	Black	Down's syndrome	-
4.	5	M	Black	-	Mild rectus exavatum.
5.	6	F	Black	-	Dysmorphic features with epicanthic folds, frontal bossing and hypotelorism. Hepatomegaly= 5cm. Short stature
6.	6	F	Black	-	Hilar adenopathy.
7.	7	F	Black	-	Not thriving very well. The calices of the left kidney look blunted on screening
8.	7	F	Black	-	Bilateral SVC's, no innominate vein. Thin, wasted child
9.	7	F	Black	-	Right supraclavicular lymph node. Poor effort tolerance, Pulmonary TB
10.	7	F	Black	-	Generalized shotty lymphadenoparthy with no obvious stigmata of immuno-suppression.
11.	10	F	Colored	-	-
12.	12	F	Black	-	-
13.	13	F	Black	-	Short stature, height below the 3rd percentile.
Cases were more than 1 type of VSD was present					
1.	12	F	B	-	-

First presentation of the VSD and the subsequent operation details

- First presentation of the VSD:

- Reason for initial VSD diagnoses
- Reason for delaying repair

Table 13: First presentation of the VSD: < 1 year age group n=9 (18%)

No	Age	Sex	Race	Reason for initial VSD diagnoses	Reason for delaying repair	Time lapse between first presentation& op
1.	0.42	M	Black	Referred for lump on occiput. Admitted at 3 weeks of age because of respiratory distress. Dysmorphic with facial asymmetry. In CCF & failure to thrive. Echo showed large inlet/outlet VSD, small PDA & pulmonary hypertension.	Waiting list	5 months
2.	0.5	F	Black	Systolic murmur heard at birth & was assessed to have small VSD. Readmitted in March 04 with bronchopneumonia, VSD re-assessed & found to be large & non restrictive. Then referred for semi urgent surgery.	Waiting list	3 months
3.	0.5	M	Black	Referred with history of tachypnoea, shortness of breath & cough. Found to have large VSD	Waiting list	3 months
4.	0.5	M	Indian	Seen shortly after birth for eye infection & cardiac murmur was heard. Assessed to have large VSD & PDA. Catheterization complicated by interrupted IVC	Waiting list	1 month

				& left side of heart could not be entered.		
5.	0.58	F	White	Assessed initially to have complete AV canal + PDA. Suspicion of membrane in LA proximal 9mm ASD secundum. 4mm PM VSD with bi-directional shunting.	Waiting list	3 months
6.	0.67	F	Black	Child had feeding problem & gastro oesophageal reflux. Found to have large malaligned VSD with severe pulmonary hypertension. Vegetations present on MV, diagnosed as SBE.	Of concern was that her sats in the ward were between 86-89% in room air.	7 months
7.	0.75	F	Black	Referred with history of recurrent chest infections, pulmonary TB, PM VSD, secundum ASD & pulmonary hypertension. Syndactyly of toes, suggestive of Aperts syndrome, no other dysmorphic features & chromosomes normal. Genetic clinic indicated probably is syndromic & is a normal child.	Waiting list	3 months
8.	0.75	F	Black	Child admitted with bronchopneumonia & CCF. Failed to thrive. Assessed to have large mid muscular VSD measuring 19.7mm on LV side, narrows to 8.9mm on RV side.	Waiting list	4 months
9.	0.83	F	Black	2 previous admissions for pneumonia, assessed to have VSD. Echo showed subarterial VSD. Prolapse of RCC into VSD but no AR. VSD in outlet septum.	Waiting list	3 months

Table 19: First presentation of the VSD: 1-4 year age group n=28 (56%)

No	Age	Sex	Race	Reason for initial VSD diagnoses	Reason for delaying repair	Time lapse between first presentation& op
1.	1	M	White	Referred with history of recurrent respiratory tract infections, failure to thrive, CCF & congenital CMV.	ASD & VSD thought to be small & would close with time.	13 months
2.	1	F	Black	Clinical features of large L to R shunt. Moderate size VSD seen on echo.	Waiting list	5 months
3.	1	M	Black	Admitted with severe bronchopneumonia. Positive Mantoux test & started on anti TB therapy. Echo showed a large PM VSD with inlet extension. In Aug 04, MPA band performed via midline incision.	Waiting list	5 months
4.	1	M	White	Child referred for systolic murmur	Large PM VSD corrected, post op residual VSD seen on echo.	3 months
5.	1	F	Black	Initially assessed to have large PDA & restrictive VSD. Admitted with lower respiratory tract infections & CFF, tachypnoea & poor feeding. Sweating & fatigue with feeds.	Waiting list	18 months
6.	1	F	Black	History of failure to thrive, palpitations, poor weight gain & cough. Assessed to have Swiss cheese VSD & pulmonary hypertension.	Surgeons decided to wait until reaches an adequate weight	18 months
7.	1	M	Black	Presented with cardiac murmur. Persistently tachypnoeic, sweating a lot & not thriving well.	Waiting list	7 months

				Echo showed abnormal systemic venous drainage, with interrupted IVC & azygos continuation. Bilateral SVC's connected by innominate vein. Interatrial septum intact. Large muscular VSD with bi-directional shunting.		
8.	1	M	Indian	Initially treated for asthma due to recurrent episodes of wheezing before he was referred for assessment of cardiac murmur. Assessed to have large 8mm malaligned outlet VSD & 3mm PDA.	Waiting list	4 months
9.	1.2	F	Black	Seen for bronchopneumonia. Incidental cardiac murmur was heard.	Waiting list	5 months
10.	1.5	F	Black	Assessed to have Down's syndrome & then later diagnosed to have TB in May 2001. Echo showed VSD`s.	Waiting list	18 months
11.	1.67	M	Black	Seen first time earlier this year, referred from GP upon finding systolic murmur. Recurrent LRTI and FTT. Echo showed large PM VSD; moderate TR; mild MR; prolapse of AMVL.	Waiting list	5 months
12.	2	F	Black	Referred by GP because of cardiac murmur. Muscular VSD 9mm, bi-directional shunting, posterior malalignment of aorta with subaortic stenosis, subaortic membrane seen. LV dilated but function good.	Waiting list	4 months
13.	2	F	Black	Admitted for bronchopneumonia & CCF. Severe failure to thrive	HIV Elisa test positive	3 months
14.	2	F	Black	Referred with history of admission for LRTI & cardiac murmur.	Waiting list	12 months
15.	2	F	Black	Referred for assessment of cardiac murmur. Found to have large sinus venosus ASD & small PM VSD. Large coronary sinus & left SVC.	Waiting list	14 months
16.	2	M	Black	Presented at 4 months of age with cardiac murmur & recurrent wheezing. Known VSD since 4 months of age. Commenced on antifailure therapy. Defaulted & admitted in March 2003 in CCF & with bronchopneumonia. 8mm PM VSD	Waiting list	14 months
17.	2	M	Black	Child has been admitted with recurrent chest infections. Echo showed large inlet/outlet VSD with bi-directional shunting. PDA could not be excluded because of severe pulmonary hypertension.	Waiting list	11 months
18.	2	M	Black	Presented with history of recurrent cough & poor weight gain. Echo & cath showed large high muscular VSD. Angiographies showed pulmonary arteries were dilated but no sudden tapering & there was prompt venous return to dilated LA.	Waiting list	5 months
19.	2	M	Black	Initially assessed to have small malaligned VSD with mild aortic regurgitation due to prolapse of an aortic cusp into VSD. DSAS was on echo.	Waiting list	4 months
20.	2	F	Black	Child was thriving well, assessed to have moderate size PM VSD with pulmonary artery pressures 60% of systemic pressures.	Waiting list	33 months
21.	2	M	Black	Referred with history of incidental murmur. No history of recurrent LRTI's, FTT or CCF	Waiting list	5 months
22.	3	F	Black	Long history of recurrent chest infection, swelling of whole body & lack of energy. On echo, large	Very high risk due to severe	21 months

				inlet/outlet VSD found, severe pulmonary hypertension. July 2002 – malnutrition & kwashiorkor. Presented in CCF with pyrexia of 38.2 degrees. Blood culture grew streptococcus viridans, treated for SBE for 6 weeks. Marked LV dysfunction	pulmonary hypertension	
23.	3	F	Indian	Referred for cardiological assessment after incidental murmur was heard. No history of effort intolerance or admissions for chest infections nor failure to thrive.	Waiting list	12 months
24.	3	M	Black	Child followed up at cardiac clinic since 4 months of age. Found to have swiss cheese VSD & small PDA & put on anti-failure therapy. Grown up reasonably well & was investigated in 2003 in view of surgery.	Waiting list	33 months
25.	3	F	Black	Child had several admissions for chest problems. Assessed to have moderate size VSD.	Waiting list	1 month
26.	4	F	Black	Presented with cardiac murmur. Echo showed double outlet RV with more than 50% override. VSD doubly committed just below aorta & measured 14mm, with bi-directional shunting across defect. Large PDA found.	Waiting list	12 months
27.	4	M	Black	Presented at age of 12 days with history of cough & blocked nose. In respiratory distress & echo confirmed diagnosis of aortic coarctation, which was repaired. MPA banding also carried out. Large muscular VSD detected.	Decided not to correct VSD at this stage, as aortic repair & MPA banding were more serious.	17 months
28.	4	F	Black	Child known to cardiac clinic since was a neonate when she was admitted with septicemia & CCF. Echo showed 5mm subaortic VSD. Prolapse of RCC into VSD with mild AR. DSAS measuring 3.3mm	Waiting list	4 months

Table 20: First presentation of the VSD: > 4 years age group n=13 (26%)

No.	Age	Sex	Race	Reason for initial VSD diagnoses	Reason for delaying repair	Time lapse between first presentation& op
1.	5	M	Black	Presented with history of swelling of feet, dyspnea & cough. Echo showed large non-restrictive malaligned PM VSD with 20% aortic override. DSAS was 4mm below aortic valve.	Waiting list	1 month
2.	5	M	Black	Presented with dyspnea, cough, malaise, history of recurrent chest infections & palpitations. Large inlet/outlet VSD.	Waiting list	6 months
3.	5	M	Black	Down's syndrome, non-restrictive VSD & small PDA.	Waiting list	1 month
4.	5	M	Black	Presented at 1 month of age with the presence of a murmur & poor feeding. Recent follow up AR was noted & PM VSD.	Waiting list	5 month
5.	6	F	Black	History of recurrent chest infections. Large muscular VSD & small PDA. Severe pulmonary hypertension & biventricular function.	High risk for surgery	9 months
6.	6	F	Black	Presented with history of cough & sore throat.	Waiting list	7 months

				Cardiac murmur heard & VSD suspected.		
7.	7	F	Black	Cath showed outlet VSD with moderate pulmonary hypertension. 30% aortic override	VSD closed off by prolapse of RCC of aortic valve into VSD.	4 years
8.	7	F	Black	Admitted for heart failure, history of recurrent chest infections & failure to thrive. Large perimembranous VSD, DSAS with no LVOTO, bilateral SVC's and no innominate vein. The left SVC was draining into the coronary sinus. She has also been suffering from recurrent LRTI's and was not growing well.	Waiting list	6 months
9.	7	F	Black	Admitted with history of cough, dyspnea & poor effort tolerance. Previously treated for TB. Echo showed large PM malaligned VSD with double outlet RV & VSD committed to the aorta.	Waiting list	1 month
10.	7	F	Black	Small malaligned VSD & DSAS. Aortic cusp prolapsing into VSD	Waiting list	5 month
11.	10	F	Colored	Seen after a murmur was incidentally found. Echo showed large PM VSD. Discrete subaortic membrane seen 10mm under aortic valve with no LVOT obstruction.	Waiting list	11 months
12.	12	F	Black	Found to have large PM outlet VSD & several smaller mid muscular VSD`s.	Waiting list	13 months
13.	13	F	Black	History of chest pain, dyspnoea and swelling of the face for 3 months. Small restrictive perimembranous malaligned VSD with right coronary cusp prolapsing into VSD & double chamber RV. Underwent cardiac catheterization for surgery.	Waiting list	

- Operation details:

- Type of procedure that was used to repair the VSD

Operation details: < 1 year age group n=9 (18%)

No	Age	Sex	Race	Type of procedure that was used to repair the VSD
1.	0.42	M	Black	Chords crossing VSD were detached from ventricular septum. VSD closed with vascular goretex patch. PDA ligation.
2.	0.5	F	Black	PFO surgically created & LV entered via PFO & LA.VSD closed with goretex patch.
3.	0.5	M	Black	Closure of large PM VSD & PFO with patch of bovine pericardium
4.	0.5	M	Indian	PDA ligation. Closure of VSD with goretex vascular patch.
5.	0.58	F	White	PDA ligation, closure of PM VSD with bovine pericardial patch, unroofing of coronary sinus into LA + snare placed around left SVC, septation of atrium with bovine pericardial patch. Died on 22 March 2004 due to presence of large amounts of serous fluids in both pleural cavities.
6.	0.67	F	Black	Closure of VSD with patch of bovine pericardium. Chord crossing VSD detached at level of edge of VSD. Sternum left open because of hyperinflated lungs.
7.	0.75	F	Black	VSD closed with vascular goretex patch. ASD also closed.
8.	0.75	F	Black	RA opened, LV entered via PFO & LA. VSD appeared to be high in outlet septum. VSD closed through right ventriculotomy with patch of bovine pericardium
9.	0.83	F	Black	Right ventriculotomy performed but was too near to septum & had to be closed. 2nd ventriculotomy performed lower on ventricle & gave perfect exposure. VSD closed with goretex vascular patch.

Operation details: 1-4 year age group n=28 (56%)

No	Age	Sex	Race	Type of procedure that was used to repair the VSD
1.	1	M	White	VSD closed with vascular goretex patch & ASD closed
2.	1	F	Black	PM VSD closed with patch of vascular goretex. Small muscular VSD closed with single pledgetted suture of Prolene 5/0.
3.	1	M	Black	LV entered via PFO & LA. Closure of VSD with patch of bovine pericardium. Removal of MPA band from around the MPA.
4.	1	M	White	Closed with a bovine patch.
5.	1	F	Black	VSD closed with patch of vascular goretex.
6.	1	F	Black	Closure PM VSD + mid muscular VSD. TV retracted to expose VSD. PM VSD closed with vascular goretex patch. Mid muscular VSD closed between 2 strips of goretex. Use of MPA band discussed, decided against due to drop of PA pressures hoping she will outgrow the residual VSD`s.
7.	1	M	Black	Closure of large muscular VSD with vascular goretex patch, ligation of left SVC.
8.	1	M	Indian	Closure of VSD with vascular goretex patch. PDA ligation
9.	1.2	F	Black	Closed with vascular goretex patch.
10.	1.5	F	Black	PDA ligation within pulmonary artery, Closure of PM VSD & mid muscular VSD with separate goretex patch, Closure of antero septal commissure of TV with one stitch.
11.	1.67	M	Black	PFO created & LA entered. PTFE patch then secured to close VSD.
12.	2	F	Black	PFO surgically created & LV entered via PFO & LA. Resection of subaortic membrane. Subarterial VSD closed via right ventriculotomy, using patch of vascular goretex. Pulmonary annulus used to secure patch superiorly.
13.	2	F	Black	Closed with patch of vascular goretex.
14.	2	F	Black	Patch closure of VSD using bovine pericardium
15.	2	F	Black	Closure of PM VSD with vascular goretex patch, Exploration of PV, Closure of secundum ASD with bovine pericardial patch, Limited modified tricuspid annuloplasty on double orifice tricuspid valve
16.	2	M	Black	PFO created & LA entered. PTFE patch then secured to close VSD.
17.	2	M	Black	Chords crossing VSD detached at level of VSD crest. VSD closed with vascular goretex patch. PDA ligation.
18.	2	M	Black	VSD closed with Goretex patch
19.	2	M	Black	Resection of DSAS. AV repair. Closure of PM VSD with Goretex patch
20.	2	F	Black	Chords crossing VSD detached from free edge of VSD. VSD closed with patch of vascular goretex.
21.	2	M	Black	Closure of PM VSD using vascular goretex patch & inspection of subaortic area.
22.	3	F	Black	TV retracted to expose VSD. Chords crossing VSD detached from inferior edge of VSD. VSD closed with vascular goretex patch.
23.	3	F	Indian	VSD closed with Goretex patch
24.	3	M	Black	PDA ligation. TV retracted to expose VSD. Antero septal chords detached. Subaortic membrane resected through VSD. Closure of VSD goretex vascular patch. Cords reattached to VSD patch inferiorly.
25.	3	F	Black	VSD closed directly using the thick fibrous tissue surrounding it.
26.	4	F	Black	Closure of VSD with vascular goretex patch. PDA ligation
27.	4	M	Black	Closure of muscular VSD with patch of vascular goretex, Removal of MPA band, Widening of MPA with patch of bovine pericardium
28.	4	F	Black	Direct closure of small VSD with single suture of Prolene. Inspection of AV.

Operation details: > 4 years age group n=13 (26%)

No.	Age	Sex	Race	Type of procedure that was used to repair the VSD
1.	5	M	Black	Chords crossing VSD detached from free edge of VSD. Fibrous membrane attached to aortic cusp was resected. VSD closed with goretex vascular patch.
2.	5	M	Black	Closure of VSD with patch of vascular goretex.
3.	5	M	Black	Closure of VSD with vascular goretex patch, PDA ligation

4.	5	M	Black	Closure of VSD using goretex patch & resection of sub aortic membrane.
5.	6	F	Black	Closure of VSD with patch of vascular goretex.
6.	6	F	Black	Closure of inlet VSD with vascular goretex patch. Mitral valve repair: closure of cleft of AML. Closure ASD secundum
7.	7	F	Black	VSD closed with vascular goretex patch.
8.	7	F	Black	Resection of DSAS. PFO surgically created & LV entered via PFO & LA. Large drain placed into coronary sinus to drain left SVC blood. VSD closed with patch of vascular goretex.
9.	7	F	Black	PFO created surgically & LV drained through PFO & LA. Atrial septum widely opened to expose MV. Supra-mitral membrane identified & resected from posterior annulus. As it was attached to AML it was carefully but only partially shaved of. VSD closed with patch of bovine pericardium. Left a 4mm PFO open.
10.	7	F	Black	Closure of VSD with patch of vascular goretex. Ligation of small left SVC
11.	10	F	Colored	Closure VSD with vascular goretex patch, Resection DSAS
12.	12	F	Black	Goretex patch closure of large muscular PM outlet VSD. Mid muscular VSD`s were a problem because of presence of large papillary muscle. These were closed with pledgetted sutures.
13.	13	F	Black	PFO was surgically created & LV entered via PFO & LA. VSD closed with patch of vascular goretex.

VSDs at operation

Table 21: Types & sizes of the VSDs: Total sample n=45 (100%)

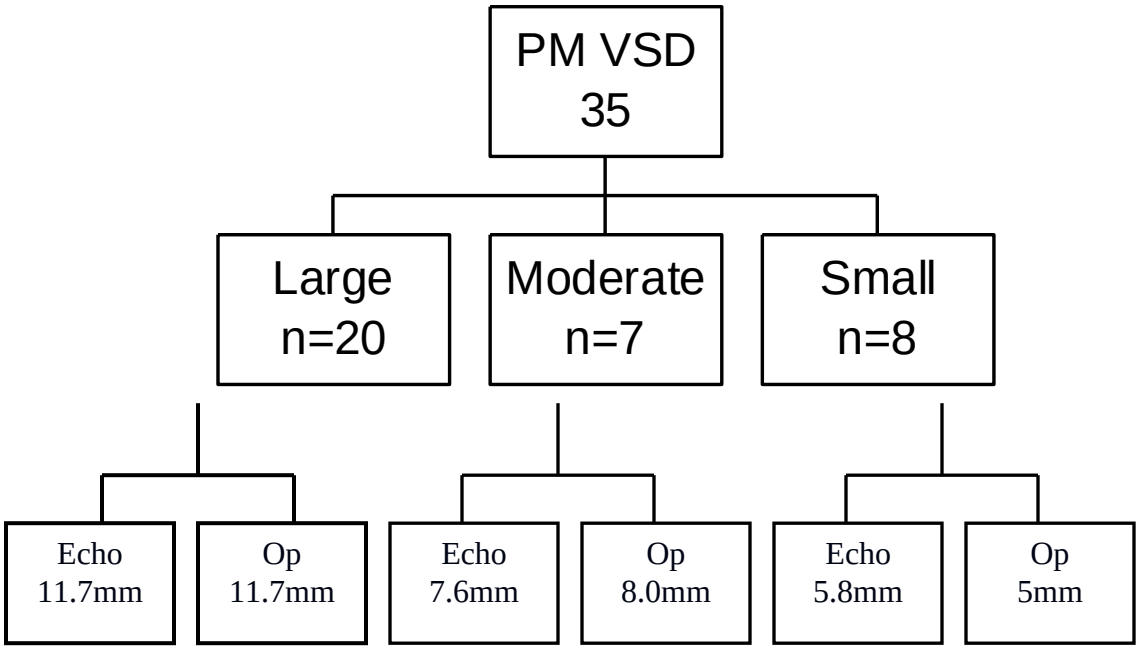


Table 21: Types & sizes of the VSDs: Total sample n=45 (100%)

	Muscular n=6	Outlet n=3	Inlet n=1
Echo (mm)	9.3mm	10.3mm	9.0mm
Operation (mm)	10.2mm	11.0mm	7.0mm

Table 22: Types & sizes of the VSDs: < 1 year age group n=8 (17.8%)

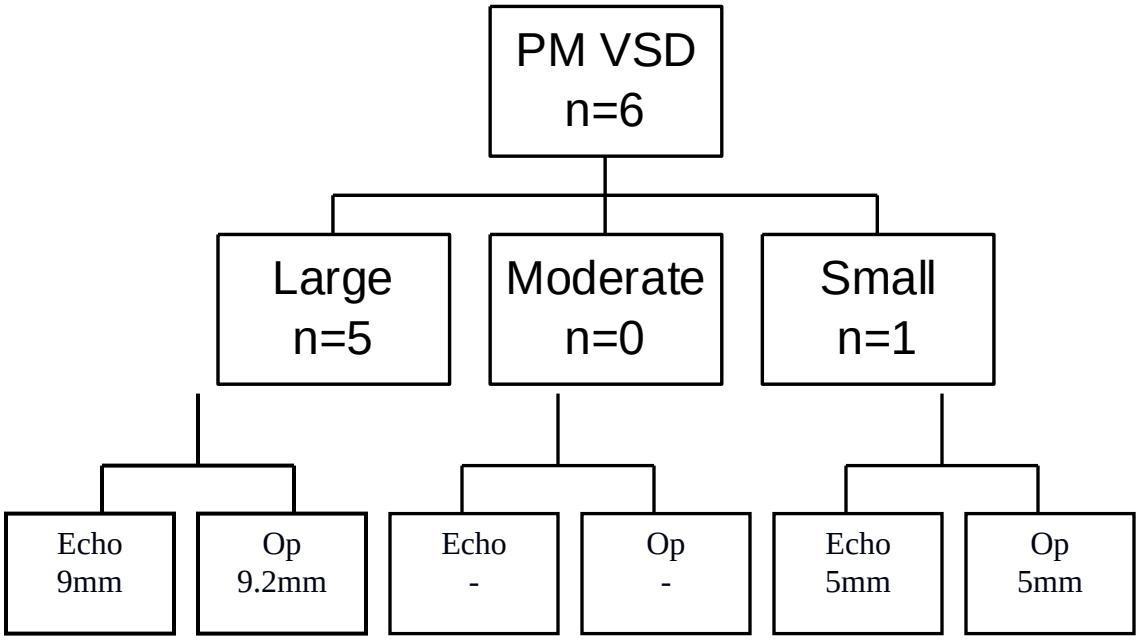


Table 22: Types & sizes of the VSDs: < 1 year age group n=8 (17.8%)

	Muscular n=1	Outlet n=1	Inlet n=0
Echo (mm)	6	7	-
Operation (mm)	6	8	-

Table 23: Types & sizes of the VSDs: 1-4 year age group n=25 (55.6%)

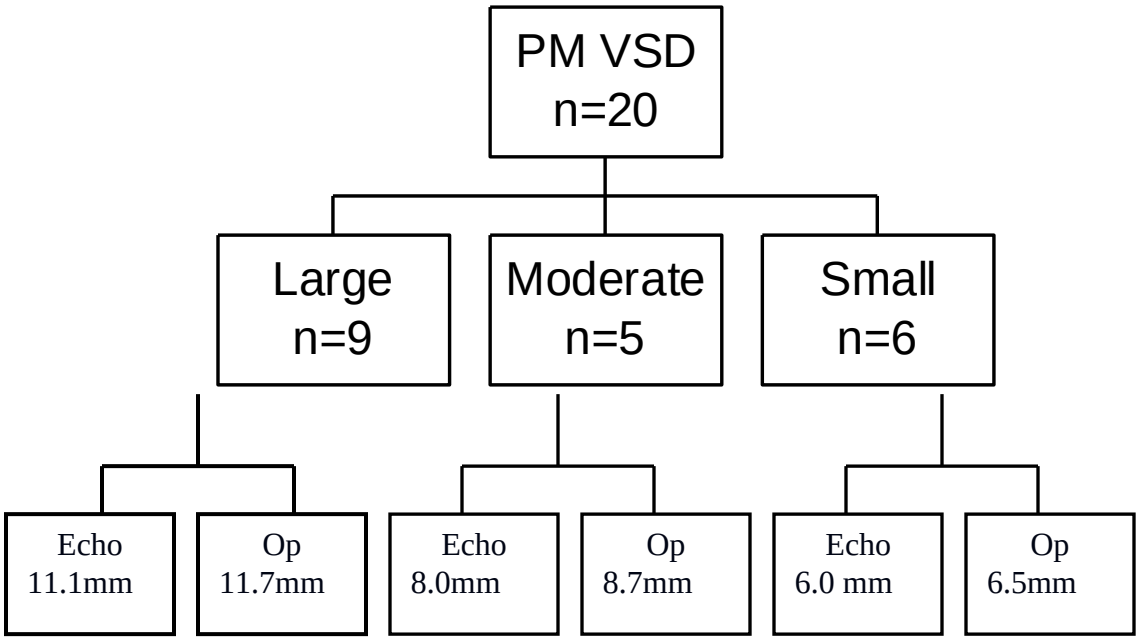


Table 23: Types & sizes of the VSDs: 1-4 year age group n=25 (55.6%)

	Muscular n=3	Outlet n=1	Inlet n=1
Echo (mm)	9.7	14	9
Operation (mm)	10.7	15	7

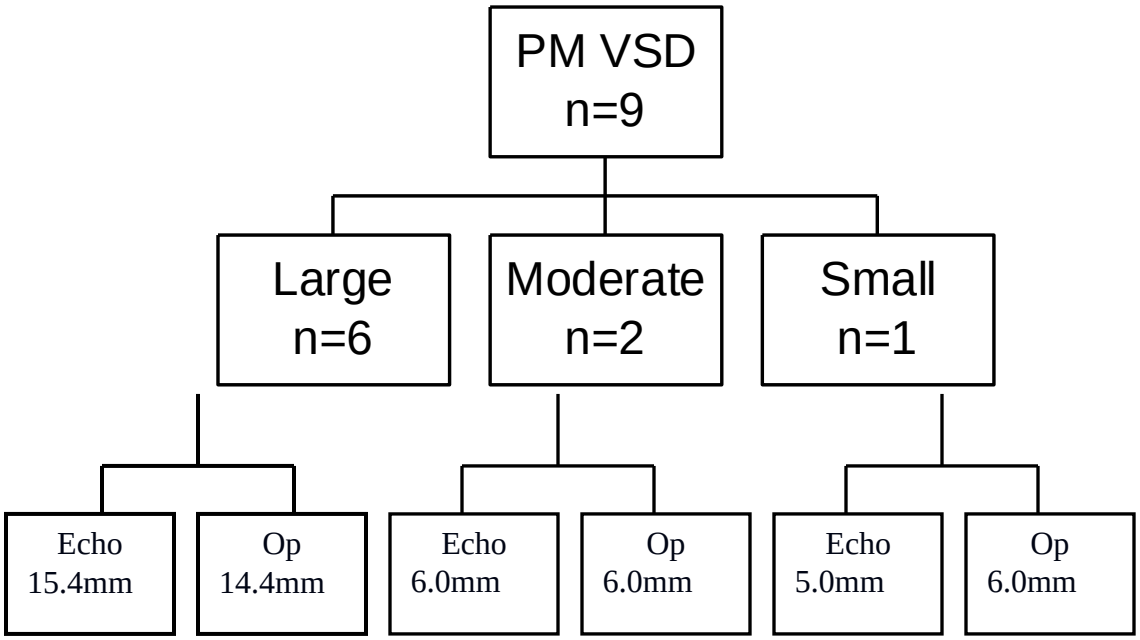


Table 24: Types & sizes of the VSDs: > 4 years age group n=13 (26%)

	Muscular n=2	Outlet n=1	Inlet n=0
Echo (mm)	10.5	10	-
Operation (mm)	11.5	10	-

Location & detailed description of the VSD in the inter ventricular septum: Perimembranous VSDs n = 35

Large PM n=20

No	Age	Sex	Race	**Location & description of VSD- Large Perimembranous n=20**
1.	0.42	M	Black	Large PM VSD with inlet extension. Chords from septal leaflet crossing VSD.
2.	0.5	F	Black	VSD in PM area
3.	0.5	M	Black	VSD in PM position.
4.	0.5	M	Indian	Septal commissural chords crossing VSD
5.	0.67	F	Black	Commissural chord crossing VSD.
6.	1	M	Black	VSD in PM area extending into inlet septum. Bar of muscle present between AV & antero-septal commissure of TV.
7.	1.67	M	Black	Dextrorotation of heart, large PM VSD with inlet extension; thickened anterior tricuspid leaflet.

8.	2	F	Black	Large PM malaligned VSD. Commissural chord of TV crosses VSD.
9.	2	F	Black	-
10.	2	M	Black	Prolapsing NCC, fibrous tissue at postero inferior edge of VSD.
11.	2	M	Black	Large PM VSD with inlet extension. Commissural chords crossing VSD.
12.	2	M	Black	Large VSD in PM area. Antero septal commissural chords crossing VSD.
13.	3	F	Black	Chords from septal leaflet crosses VSD, some chords seemed to be attached to AV annulus.
14.	3	F	Indian	Large PM VSD, non restrictive
15.	5	M	Black	Large PM VSD. Chords from septal leaflet of TV cross VSD. Small superior fibrous membrane attached to lower part of sinus of aortic cusp, which prolapses into VSD. Fibrous ridge on inferior border of VSD also seen. DSAS of 3mm situated 4mm below AV.
16.	5	M	Black	VSD in PM position.
17.	6	F	Black	Chordal tissue crossing the LVOT, inserts on the crest of VSD. VSD large, inlet VSD with PM extension, partially closed off by redundant TV tissue in PM area.
18.	7	F	Black	DSAS seen on inferior edge extending onto LVOT, measured 2-3mm in width.
19.	7	F	Black	Large malaligned PM VSD with aorta arising from RV.VSD committed to aorta.
20.	10	F	Colored	Large PM VSD. Thick fibrous membrane not circumferential present low in LVOT, related to inferior edge of VSD. Partial closure of VSD by AV. Discrete subaortic membrane=6mm in length & 10mm below aortic valve. Several small echo densities attached to chordae that could be vegetations or areas of thickening of chordae.

Moderate PM n=7

No	Age	Sex	Race	**Location & description of VSD- Moderate Perimembranous n=7**
1.	1	F	Black	-
2.	1	M	Indian	VSD in PM position.
3.	2	F	Black	Edges of PM VSD fibrous, attempt by TV to close VSD, thickened antero-septal chords crossing VSD.
4.	2	M	Black	DSAS 3.5mm long & 2.72mm below AV.
5.	3	M	Black	Echo & cath showed moderate size VSD & small PDA. Discrete subaortic membrane seen but no gradient found across LVOT. VSD in PM area, but bar of muscle separated VSD from TV. Antero-septal chords crossed VSD. Thick fibrous membrane found on inferior edge of VSD on LV side. No abnormal muscle bundle in the RV.
6.	5	M	Black	PM VSD partially closed by chords from septal leaflet of TV. Discrete sub aortic membrane attached to LV side of infero-posterior margin of VSD. Membrane measures 5mm in width & 6mm below AV.
7.	7	F	Black	Malaligned PM VSD with fibrous edges, mild prolapse of non-coronary cusp into VSD. DSAS with no LVOT obstruction. Antero septal commissural chords crossing VSD.

Small PM n=8

No	Age	Sex	Race	**Location & description of VSD- Small Perimembranous n=8**
1.	0.75	F	Black	-
2.	1	M	White	Small VSD with attempted aneurismal closure by TV tissue.
3.	1.2	F	Black	PM VSD with high muscular extension. On left ventricular side of VSD small fibrous ridge present. Antero-septal commissural chords of TV thickened & fused seemed like they were attempting to close VSD.
4.	2	F	Black	Small PM VSD with fibrous edges partially closed by TV.
5.	2	M	Black	VSD in PM area. Thick subaortic membrane going from bottom of RCS to bottom of non-coronary sinus crossing the LVOT underneath the VSD. Subaortic membrane distorted the cusps & might have been primary cause of the AR. Both right and non coronary cusps were thickened & non coronary prolapsing mildly towards the LV.

6.	3	F	Black	PM VSD partially closed by TV. VSD small & elongated & surrounded by fibrous tissue. TV thickened especially in region of antero-septal commissure.
7.	4	F	Black	PM VSD surrounded by thick fibrous edges. In a subaortic position away from TV. On inspection of AV, mild prolapse of NCC & RCC found with redundancy of same cusps. Coaption of 3 cusps adequate. No DSAS but in region of VSD, small amount of fibrous tissue related to VSD.
8.	13	F	Black	VSD surrounded by fibrous edges, bar of muscle present between septal leaflet of TV &VSD. Thick fibrous tissue present on septal surface of RVOT starting at superior edge of VSD & going across RVOT. Fibrous tissue had potential to become obstructive.

Muscular VSDs n= 6

No	Age	Sex	Race	**Location & description of VSD - muscular**
1.	0.75	F	Black	VSD was high muscular towards the outlet septum & measured about 6-7mm. Difficult to find amongst RV trabeculations.
2.	1	M	Black	Large muscular VSD going down towards trabecular septum. Small bar of muscle present between TV & VSD.
3.	2	F	Black	Upper border formed by conjoined aortic & pulmonary annuli. Inferior edge on LV side, thick fibrous membrane seen. Aortic cusp prolapsing slightly into VSD partially obstructing it.
4.	4	M	Black	VSD situated in PM region but is a muscular VSD with large bar of muscle between TV & VSD. Aorta appears to override the IVS to some degree. Membrane (PM septum) extending from aorta below AV (above the IVS) causing subaortic narrowing with opening of 7mm.
5.	5	M	Black	Large muscular VSD situated in inlet/PM area.
6.	6	F	Black	Muscular, inlet VSD with bar of muscle between VSD & TV.

Outlet VSDs n=3

No.	Age	Sex	Race	**Location & description of VSD - outlet**
1.	0.83	F	Black	True sub-arterial VSD with upper border formed by conjoint annuli of aortic & PV. Bar of muscle present between inferior border of VSD & TV. RCC of AV prolapsing into VSD practically occluding it.
2.	4	F	Black	VSD large doubly committed malaligned true subarterial just below aorta. No subaortic nor sub pulmonary conus. Hypertrophied septal muscle bundle in RV infundibular area with potential RVOT obstruction.
3.	7	F	Black	True subarterial VSD, superior border formed by conjoint annuli of aortic & PV. Large bar of muscle between inferior borders of VSD & TV. RCC of AV prolapsing into VSD effectively closing it.

Inlet VSDs n= 1

No.	Age	Sex	Race	**Location & description of VSD - inlet**
1.	1	M	White	Residual VSD was in inlet septum close to postero-septal commissure of TV. Long narrow VSD with muscle band crossing it. Separated from TV by small bar of muscle.

Table 25: Types, sizes, location & detailed description of the VSD in the inter ventricular septum: cases where more than one type of VSD occurred n=5 (10%)

No.	Age	Sex	Race	Types of VSD`s	Size of VSD (mm)		Location & description of VSD
					Echo	Op	

1.	0.58	F	White	PM & apical muscular	4	4	-
2.	1	F	Black	PM & muscular	PM: 10 M: 6	PM: 13.5 M: 6	PM VSD malaligned, prolapse of aortic cusp into VSD partially occluding it. 2 residual muscular VSD`s seen at apical region. Largest at apex= 9mm and smallest-5mm (echo).
3.	1	F	Black	PM, mid muscular & Swiss cheese	-	PM: 5 M: 2	1 VSD in PM area, some attempt by TV to close VSD. Edges of VSD fibrous. Another small muscular VSD situated inferiorly under infero-septal commissure in muscular septum.
4.	1.5	F	Black	PM & mid muscular	M: 7	M: 7	2 VSD`s: small PM VSD & larger moderate sized high muscular VSD.
5.	12	F	Black	PM & 2 mid muscular	PM: 8.5	PM: 12	PM VSD separated from TV by large muscle band. Middle of trabecular septum, large papillary muscle to which chords to anterior tricuspid leaflet were attached. Either side of large papillary muscle slit like VSD`s, longer on left side of papillary muscle.

Appearance of the VSDs when viewed from the different chambers of the heart: perimembranous VSDs n=35

Tables 26 – 29

Large PM n=20

Appearance of VSD- Large Perimembranous n=20

No	Age	Sex	Race	RV	LV 70	LV 40	Other
1.	0.42	M	Black	-	-	-	-
2.	0.5	F	Black	-	-	-	-
3.	0.5	M	Black	-	-	-	-
4.	0.5	M	Indian	-	-	-	-
5.	0.67	F	Black	-	-	-	-
6.	1	M	Black	-	-	-	-
7.	1.67	M	Black	-	RV trabeculated & dilated. VSD measures 12mm. Coronary arteries seen, appear normal.	Good contractility of LV. No obvious evidence of non compaction	Angiogram shows good size PA branches. Good peripheral aborization on left lung but right lung appears to have decreased hazy background suggestive of probable vascular disease.
8.	2	F	Black	-	Moderate to large PM VSD.	-	-
9.	2	F	Black	-	Good LV contractility. Contrast seen ejecting across VSD into RV. Bulge seen in R sinus. RCC seen filling.	-	Mild aneurismal dilatation of right sinus of valsalva seen & no shunting into RA or RV seen. RCC seen. Normal head &

							neck vessels.
10.	2	M	Black	-	Enlarged LV outlined. Simultaneous filling of ascending aorta & arch. Moderate sized PM VSD seen filling an enlarged RV	-	Aortogram shows left aortic arch. Origins of head & neck vessels normal.
11.	2	M	Black	-	Good LV contractility. Contrast seen ejecting from RV through fenestrated VSD into RV. R to L shunting & prompt venous return to LA.	Shows a trabeculated RV & dilated PA lying anterior to & arising from RV.	-
12.	2	M	Black	-	Large muscular VSD. Contrast seen filling RV from LV. LV dilated, has good contractility. MPA dilated but no RVOT obstruction	-	Ascending aortogram shows coronary arteries appear normal, no coarctation of aorta, left sided aortic arch
13.	3	F	Black	-	Shows a left to right shunt across VSD to RV	L to R shunting. LV contractility not normal. Dominant LCA system fills from aorta. RCA not well seen.	-
14.	3	F	Indian	-	LV dilated. Moderate size PM VSD with L to R shunting across it. RV slightly dilated. Left sided aortic arch, normal branches.	-	No PDA present
15.	5	M	Black	-	VSD well seen with L to R shunting across it. Ascending aorta & its branches are normal.	-	Ascending aortogram done to exclude PDA. Ductal bump seen but no PDA.
16.	5	M	Black	-	-	-	-
17.	6	F	Black	-	Good size LV contracting well. Large inlet VSD. No gooseneck deformity.	VSD not seen very well in this view.	-
18.	7	F	Black	-	-	-	Large VSD present with L to R shunting across it. LVOT is normal
19.	7	F	Black	RV tri-partite ventricle & is dilated. Aorta & PAs fill simultaneously confirming double outlet anatomy.	LV not dilated. Large VSD with L to R shunting across it. Aorta fills immediately & is therefore committed to VSD.	Sub aortic tunnel. Aortic & PV appear to be at same level, great vessels side by side.	PA`s good size, no evidence of stenosis. Left upper PV crosses over & enters LA on the right.
20.	10	F	Colored	-	-	-	-

Moderate PM n=7

Appearance of VSD- Moderate Perimembranous n=7							
No	Age	Sex	Race	RV	LV 70	LV 40	Other
1.	1	F	Black	-	Good contractility of heart. RV fills through L to R shunt across VSD. Prompt venous return to dilated LA.	-	Descending aortogram shows that PDA been successfully ligated.
2.	1	M	Indian	-	Enlarged LV outlined. Simultaneous filling of ascending aorta & arch noted. Moderate size PM VSD seen filling enlarged RV. No LVOT obstruction.	-	Aortogram shows normal L aortic arch including descending portion. Origin of head & neck vessels normal. No aortic regurgitation noted. Small PDA of 3mm. Normal L & R coronary arteries & branches.
3.	2	F	Black	-	VSD with L to R shunt seen. PA`s fill from RV. Normal coronary arteries.	-	-
4.	2	M	Black	-	Moderately sized PM VSD with L to R shunting across VSD.	-	-
5.	3	M	Black	RV normal size. Inlet, trabecular & outlet portions well visualized. Appears to be narrowing of infundibular area during systole.	Moderately sized PM VSD. Contrast seen filling RV from LV across defect. Both ventricles appear normal. No coarctation of aorta.	-	Descending aortogram shows tiny PDA. Contrast seen filling MPA from descending aorta across defect. Ampulla of PDA appears very small. Descending & ascending aorta appears normal.
6.	5	M	Black	-	Small PM VSD seen. Contrast seen filling RV from LV. Cranial vessels seen coming of arch of aorta normally. No coarctation of aorta.	-	Ascending aortogram shows mild AR. Coronary arteries appear normal. No coarctation of aorta.
7.	7	F	Black	-	Small PM VSD. Left aortic arch, no coarctation. Cranial vessels come off aortic arch normally.	-	Ascending aortogram shows trivial AR. Normal coronary arteries. Left sided aortic arch, no coarctation. Normal cranial vessels.

Small PM n=8

Appearance of VSD- Small Perimembranous n=8							
No	Age	Sex	Race	RV	LV 70	LV 40	Other
1.	0.75	F	Black	-	-	-	-

2.	1	M	White	-	Moderately sized PM VSD. Contrast crosses from LV to RV across VSD. Both ventricles of normal size. Left sided aortic arch.	-	-
3.	1.2	F	Black	-	Moderate size PM VSD. RV seen filling across VSD from L to R shunt. Large PA seen filling.	-	Ascending aortogram shows normal aorta & head & neck vessels. Normal coronary arteries. No PDA or coarctation of aorta.
4.	2	F	Black	-	-	-	-
5.	2	M	Black	-	LV well outlined & appears normal in size. LVOT well defined & no obstruction noted. Small VSD noted filling RV. LA is normal. Left sided aortic arch seen.		Aortogram shows normal L sided aortic arch including descending portion. Origins of head & neck vessels noted. Normal L & R coronary artery branches. Moderate AR noted filling both RV & LV.
6.	3	F	Black	-	Moderately sized PM VSD. Contrast crosses defect from LV to RV. Ventricles of good size with good contractility. L aortic arch with no coarctation.	-	-
7.	4	F	Black	-	LV not dilated. L aortic arch with normal branching. Tiny sub aortic VSD with L to R shunting across it.	-	-
8.	13	F	Black	Small PM-VSD. Aorta normal size. No coarctation of aorta & cranial vessels appear normal	Good size LV, good contractility. Small PM-VSD, aorta appears normal.	-	-

Muscular VSDs n= 6

Appearance of VSD- muscular							
No	Age	Sex	Race	RV	LV 70	LV 40	Other
1.	0.75	F	Black	-	A large mid muscular VSD is seen with a L to R shunt. VSD measures 19.7mm on LV side & narrows to 8.9mm on RV side. Mitral regurgitation. No PDA.	-	-
2.	1	M	Black	-	Normal L aortic arch & dilated LV. High muscular VSD seen	-	-

					with L to R shunt. PA`s seen filling from RV.		
3.	2	F	Black	Good size RV with inlet, outlet & trabecular parts well visualized. Good contractility of ventricle & dilatation of RVOT.	VSD in outlet position, cranial vessels come off L aortic arch normally.	Outlet VSD visible, L aortic arch & no coarctation of aorta, cranial vessels come off arch normally.	-
4.	4	M	Black	-	-	-	-
5.	5	M	Black	-	-	-	-
6.	6	F	Black	-	Large muscular VSD with L to R shunt. Both ventricles enlarged. LV contractility appears reduced. Normal L aortic arch.	-	Descending aortogram shows a small PDA shunting L to R.

Outlet VSDs n= 3

Appearance of VSD- Outlet							
No	Age	Sex	Race	RV	LV 70	LV 40	Other
1.	0.83	F	Black	-	Moderate size VSD in sub arterial position with L to R shunting.	-	-
2.	4	F	Black	RV appears small & tripartite. Large MPA segment. Pulmonary venous return to dilated LA & normal sized LV.	LV normal size with good contractility	-	Descending aortogram shows large PDA.
3.	7	F	Black	-	Moderate sized outlet VSD just below AV. Contrast seen filling RV across VSD. Big MPA segment visualized.	-	-

Inlet VSDs n= 1

Appearance of VSD- Inlet							
No	Age	Sex	Race	RV	LV 70	LV 40	Other
1.	1	M	White	-	-	-	-

Cases were more than 1 type of VSD

Appearance of VSD- > 1 type of VSD							
No	Age	Sex	Race	RV	LV 70	LV 40	Other
1.	0.58	F	White	-	Small LV outlined. Simultaneous filling of ascending aorta & arch noted.	-	-

					Normal origins of head & neck vessels noted.		
2.	1	F	Black	-	Shows residual VSD, which is high up in PM region with L to R shunting.	-	LPA angiogram shows dilated MPA & peripheral branches that are thickened & torturous.
3.	1	F	Black	-	Shows reasonable size LV, contracting well. Head & neck vessels normal.	Anomalous origin of head & neck vessels.	-
4.	1.5	F	Black	-	LV is of good size & good contractility.	-	-
5.	12	F	Black	-	Shows clearly single large VSD in high muscular area & 2 smaller VSDs in mid muscular area.	-	-

Great vessels of the heart

Tables 30-40

Aorta and valve abnormalities: < 1 year age group n=9 (18%)

No.	**Age**	**Sex**	**Race**	**Ascend part (mm)**	**Arch**			**Valves (mm)**				**Descend part (mm)**
					R/L	*% override*	*Size (mm)*	*AV*	*MV*	*TV*	*Details*	
1.	0.42	M	Black	10	Left	-	-	7	-	-	-	11
2.	0.5	F	Black	7	Left	-	-	7.3	-	-	-	-
3.	0.5	M	Black	10	Left	-	-	TV annulus small anterior leaflet prolapsing				6
4.	0.5	M	Indian	-	Left	25	-	13	-	-	-	-
5.	0.58	F	White	9	Left	-	-	9	12	18	-	-
6.	0.67	F	Black	-	Left	17.5	-	-	-	-	Severe TR	8
7.	0.75	F	Black	-	Left	-	11	-	-	-	-	-
8.	0.75	F	Black	-	Left	-	-	8.8	17	-	Mild TR	9
9.	0.83	F	Black	8	Left	20	11	-	18	11	RCC of AV prolapsing into VSD practically occluding it.	-

Aorta and valve abnormalities: 1-4 year age group n=28 (56%)

No.	**Age**	**Sex**	**Race**	**Ascend part (mm)**	**Arch**			**Valves (mm)**				Descend part (mm)
					R/L	*% override*	*Size (mm)*	*AV*	*MV*	*TV*	*Details*	
1.	1	M	White	-	Left	-	11	-	-	-	-	-
2.	1	F	Black	-	Left	-	-	-	-	-	-	-
3.	1	M	Black	-	Left	7.5	-	Mild TR				-
4.	1	M	White	-	Left	-	-	12	18	23	-	5.7
5.	1	F	Black	-	Left	30	-	9	-	-	-	-
6.	1	F	Black	Dilated	Left	50	-	-	24	21	Mild M + T R	-

7.	1	M	Black	12	Left	-	16	Mild TR, Tri leaflet AV.				-
8.	1	M	Indian	8	Left	5	17	-	-	-	-	-
9.	1.2	F	Black	13.3	Left	-	-	-	-	-	-	6.6
10.	1.5	F	Black	-	Left	-	-	-	-	-	-	-
11.	1.67	M	Black	-	-	40	-	Moderate TR, Mild MR. Prolapse of AMVL.				-
12.	2	F	Black	8.5	Left	-	12	5	-	-	PM septum jutting out below AV causing tunnel effect.	6.6
13.	2	F	Black	17.4	Left	30	-	-	-	-	-	-
14.	2	F	Black	-	Left	-	-	12	-	-	-	-
15.	2	F	Black	-	Left	-	-	TV had 2 orifices, inferior half the size of superior orifice. 2 orifices separated by fibrous band to which leaflet tissue attached. 4 leaflets identified on larger orifice, 3 on smaller orifices, leaflets failed to coapt.				6
16.	2	M	Black	-	Left	25	16	-	20	16	RCC of AV prolapsing into VSD	-
17.	2	M	Black	Small	-	-	-	11	-	-	-	-
18.	2	M	Black	-	Left	-	-	-	-	-	-	16
19.	2	M	Black	18	Left	-	20	17	-	-	Thickened tips of AV leaflets. Mild AR.	-
20.	2	F	Black	-	-	-	-	-	-	-	-	-
21.	2	M	Black	-	Left	-	16	16	21	20	-	-
22.	3	F	Black	14	Left	-	-	13.8	-	-	-	-
23.	3	F	Indian	14	Left	-	-	12	20	18	-	12
24.	3	M	Black	13	Left	-	-	-	-	-	-	12
25.	3	F	Black	-	Left	-	12	-	15	12	-	-
26.	4	F	Black	Small, right & posterior to PA.	Left	50	19	14	18	15	-	Pulsatile
27.	4	M	Black	-	Left	-	-	16	14	15	AV abnormal, bicuspid, opening = 4.4mm.	7.6
28.	4	F	Black	-	Left	20	15	14	17	17	Mild AR.	-

Aorta and valve abnormalities: > 4 years age group n=13 (26%)

No	**Age**	**Sex**	**Race**	**Ascend part (mm)**	**Arch**			**Valves (mm)**				**Descend part (mm)**
					R/L	*% override*	*Size (mm)*	*AV*	*MV*	*TV*	*Details*	
1.	5	M	Black	25	Left	20	-	-	-	-	-	7
2.	5	M	Black	-	-	-	-	-	-	-	-	-
3.	5	M	Black	-	-	-	-	-	-	-	-	-
4.	5	M	Black	23	Left	-	-	-	-	-	AV cusp prolapsing into VSD. Mild AR.	-
5.	6	F	Black	16	Left	-	-	-	19	25	Mild TR.	8

6.	6	F	Black	13	Left	-	-	MV not AV canal type of valve. Large posterior leaflet occupying 2/3 of annular circumference, anterior MV leaflet large true cleft (9.6mm) whole way up to annulus, dividing AML into large antero-lateral smaller postero-medial portions.				-
7.	7	F	Black	16.2	Left	-	17.4	RCC of AV prolapsing into VSD effectively closing it.				-
8.	7	F	Black	Good size	Left	-	-	15	24	21	DSAS 4.1mm 10 mm below AV	-
9.	7	F	Black	-	Left	-	-	21	18	23	Sub aortic area 7mm potential for sub aortic stenosis post VSD repair.	-
10.	7	F	Black	16.4	Left	30	-	Prolapse of RCC & NCC into VSD.				7
11.	10	F	Colored	-	Left	-	-	-	-	-	-	-
12.	12	F	Black	-	-	-	-	17	-	-	-	-
13.	13	F	Black	-	Left	25	19	RCC of AV prolapsing within VSD occluding it.				-

Pulmonary vessels: < 1 year age group n=9 (18%)

No.	**Age**	**Sex**	**Race**	**Pulmonary Trunk (mm)**		**Pulmonary Valve (mm)**		**Pulmonary Artery (mm)**			
				Size	*Description*	*Size*	*Description*	*MPA*	*RPA*	*LPA*	*Description*
1.	0.42	M	Black	-	-	-	-	16.5	-	9	MPA dilated
2.	0.5	F	Black	-	-	-	-	12.5	7	8	MPA dilated
3.	0.5	M	Black	-	-	-	-	15	-	10	MPA dilated, proximal part aneurismal & fragile
4.	0.5	M	Indian	-	-	14.6	-	15.5	9	8	MPA large & tense
5.	0.58	F	White	-	-	15	-	19	-	-	MPA dilated
6.	0.67	F	Black	-	-	10	-	12	6	6	MPA dilated to twice size of aorta.
7.	0.75	F	Black	-	-	-	-	18	11	10	-
8.	0.75	F	Black	-	-	16	-	16	10	9	-
9.	0.83	F	Black	-	-	10	-	15	7	6	-

Pulmonary vessels: 1-4 year age group n=28 (56%)

No.	**Age**	**Sex**	**Race**	**Pulmonary Trunk (mm)**		**Pulmonary Valve (mm)**		**Pulmonary Artery (mm)**			
				Size	Description	Size	Description	MPA	RPA	LPA	Description
1.	1	M	White	-	-	-	-	-	-	-	-
2.	1	F	Black	-	-	-	-	-	-	-	-
3.	1	M	Black	-	Bilateral bronchopneumonic changes	-	-	-	-	-	-
4.	1	M	White	-	-	13	-	15	6	8	-
5.	1	F	Black	-	-	-	-	PA enlarged. MPA dilated. Lung field plethoric, no evidence of peripheral pruning.			
6.	1	F	Black	-	-	17	-	16	8	9	MPA+ PA

											dilated with mild PR.
7.	1	M	Black	-	-	-	-	19	7	8	MPA + PAs dilated & tortuous.
8.	1	M	Indian	-	Slight tortuosity of vessels. No tapering noted.	-	-	15	-	-	MPA dilated & tense
9.	1.2	F	Black	-	-	-	-	-	-	-	-
10.	1.5	F	Black	-	-	-	-	-	-	-	-
11.	1.67	M	Black	-	-	18	-	18	9	11	-
12.	2	F	Black	-	-	-	Mild PR, no valvar stenosis	18	-	-	-
13.	2	F	Black	-	-	-	-	18.3	-	-	-
14.	2	F	Black	-	-	15	-	16	12	11	-
15.	2	F	Black	-	PVs all drained on left.	-	-	15	-	-	-
16.	2	M	Black	-	-	-	-	-	-	-	MPA dilated
17.	2	M	Black	-	-	-	-	20	10	8	-
18.	2	M	Black	-	-	-	-	26.4	-	-	PA`s appear dilated, no sudden tapering
19.	2	M	Black	-	-	14	-	12	7	7	-
20.	2	F	Black	-	-	-	-	-	-	-	-
21.	2	M	Black	-	-	-	-	16.4	9	9.5	-
22.	3	F	Black	-	-	-	-	15.5	-	-	RPA has large branching tortuous proximal vessel.
23.	3	F	Indian	-	-	22	-	22	9	13	-
24.	3	M	Black	-	-	-	-	14	-	-	PA large & tense.
25.	3	F	Black	-	-	-	-	18	7	7	-
26.	4	F	Black	-	-	17	-	20	11	11.6	MPA massive, ductal bump
27.	4	M	Black	-	-	-	-	11	6.6	6.7	Dense adhesions in region of MPA band. Band tight, impinging on RPA.
28.	4	F	Black	-	-	10	-	11	4	5	-

Pulmonary vessels: > 4 years age group n=13 (26%)

No.	**Age**	**Sex**	**Race**	**Pulmonary Trunk (mm)**		**Pulmonary Valve (mm)**		**Pulmonary Artery (mm)**			
				Size	Description	Size	Description	MPA	RPA	LPA	Description
1.	5	M	Black	-	-	-	-	20	15	11	-
2.	5	M	Black	-	-	-	-	-	-	-	-
3.	5	M	Black	-	-	-	-	-	-	-	PA dilated
4.	5	M	Black	-	-	-	-	-	-	-	-
5.	6	F	Black	-		25	Mild PR.	28	10	9	MPA very large & tense

6.	6	F	Black	-	-	-	-	-	-	-	-
7.	7	F	Black	-	-	-	-	23.4	-	-	Big PAs noted.
8.	7	F	Black	-	-	22	-	16	11	11	-
9.	7	F	Black	-	-	16	Poor mobility of PMVL	15	9	11	Peripheral tapering in keeping with severe PHT.
10.	7	F	Black	-	-	-	-	-	-	-	-
11.	10	F	Colored	-	-	-	-	-	-	-	-
12.	12	F	Black	-	-	-	-	30	-	-	MPA large & tense
13.	13	F	Black	-	-	-	-	17	11	13	-

Superior vena cava, inferior vena cava & associated structures

No	**Age**	**Sex**	**Race**	**Superior Vena Cava (mm)**		**Inferior Vena Cava (mm)**	
				Size	Description	Size	Description
1.	0.5	M	Indian	-	-	-	Interruption of IVC with azygos continuation to SVC & RA.
2.	0.58	F	White	-	Bilateral SVCs, L SVC drained into large coronary sinus. Both SVCs similar in size, L SVC marginally smaller. Dilated R SVC. No innominate vein. Branch from hemi-azygos noted draining into SVC.	-	-
3.	1	M	White	-	L SVC draining into LA with unroofed coronary sinus	-	-
4.	1	M	Black	R=12 L=5	R SVC large drains normally into RA. L SVC draining into roof of LA next to L atrial appendage or into unroofed coronary sinus (5.2mm). Bilateral SVCs connected by small innominate vein.	-	Interrupted IVC with azygos continuation drains into RA superiorly.
5.	1.2	F	Black	-	-	3.9	-
6.	1.67	M	Black	-	Suspected bilateral SVCs with rudimentary innominate vein. No innominate seen filling therefore L SVC suspected.	-	-
7.	2	F	Black	-	-	5	-
8.	3	F	Black	-	Bilateral SVCs with no innominate vein. SVCs drain into a large coronary sinus of 7mm.	-	Large and dilated
9.	4	M	Black	-	-	8	Patent supra hepatic IVC
10.	6	F	Black	-	-	11	Dilated
11.	7	F	Black	L=6	Bilateral SVC's, no innominate vein. L slightly smaller than R, L drained into coronary sinus (6mm by 13mm)	-	-
12.	7	F	Black	L=3.5	Left SVC small	8	-
13.	10	F	Colored	R=4 L=8	R SVC small, L large, draining into coronary sinus (8mm). No innominate vein.	-	-

Part B: VSDs occurring in TOF

1) **Demographics and general**

Table 1:

Gender distribution	***Total sample n=50***		**< 2 years n=15**		**2-4 years n=21**		**> 4 years n=14**	
	No.	*%*	No.	%	No.	%	No.	%
Males	*27*	*54*	6	40	13	61.9	8	57.1
Females	*23*	*46*	9	60	8	38.1	6	42.9

Table 2:

Population affinity	***Total sample n=50***		**< 2 years n=15**		**2-4 years n=21**		**> 4 years n=14**	
	No.	*%*	No.	%	No.	%	No.	%
Blacks	*41*	*82*	11	73.3	18	85.7	12	85.7
Whites	*3*	*6*	2	13.3	1	4.8	-	-
Indians	*1*	*2*	1	6.7	-	-	-	-
Colored	*5*	*10*	1	6.7	2	9.5	2	14.3

Table 3:

Age (yrs)	**Number**	**%**	**Minimum**	**Maximum**	**Mean**	**Standard Deviation**
Total sample	*50*	*100*	*0.67*	*13*	*3.36*	*2.48*
< 2 years	15	30	0.67	1.58	1.28	0.31
2-4 years	21	42	2	4	2.71	0.78
> 4 years	14	28	5	13	6.57	2.31

Table 4:

Weight (kg)	***Total sample n=50***	**< 2 years n=15**	**2-4 years n=21**	**> 4 years n=14**
Minimum	*6.4*	6.4	8	9

Maximum	*21.4*	11.2	16.8	21.4
Mean	*10.77*	8.63	10.88	15.24
Standard deviation	*4.53*	3.42	2.35	5.61

Table 5:

Height (m)	***Total sample n=50***	**< 2 years n=15**	**2-4 years n=21**	**> 4 years n=14**
Minimum	*0.6*	0.6	0.71	0.11
Maximum	*1.5*	0.92	1.03	1.5
Mean	*0.77*	0.76	0.84	1.00

Prenatal history

Table 6:

Prenatal history		
Normal births n= 48	*Complicated births n= 2*	
• Infants were born at term by normal vaginal delivery. • No complications.	Premature birth (n=1) • 1 year old black female • One of twins • Born to HIV + mother	Breech delivery (n=1) • 19 month old colored male • delivered at home

Previous operation details

Table 7:

Previous operation details n= 5 (10%)					
No.	*Age (yrs)*	*Sex*	*Race*	*Operation details*	*Complications*
1.	1	M	Black	Left sided Blaloch Taussig (BT) shunt with a 4mm goretex thin walled graft was inserted as a life saving procedure in order to reduce cyanosis.	No complications

2.	4	M	Black	a) Tracheo-oesophageal fistula repair b) Central shunt c) RVOT patch	• infant developed metabolic acidosis & Klebsiella sepsis • Central shunt failed • Patch thickened & calcified in places.
3.	4	M	White	BT shunt	Shunt became stenosed
4.	5	M	Black	a) Closure of VSD b) Widening of RVOT,MPA & the origin of the LPA with a bovine pericardial patch c) PV replacement d) PFO created	BT shunt that was previously inserted became stenosed
5.	9	F	Black	Right sided BT shunt	No complications

Diagnosis

VSDs: Types of VSDs and associated physiological diagnosis

Table 8:

Types of VSDs (n=50)								
	PM VSD		Muscular VSD		Outlet VSD		Inlet VSD	
	No.	%	No.	%	No.	%	No.	%
Total sample (n=50)	*44*	*88*	-	-	*6*	*12*	-	-
< 2 years (n=15)	11	73.3	-	-	4	26.7	-	-
2-4 years (n=21)	21	100	-	-	-	-	-	-
> 4 years (n=14)	12	85.7	-	-	2	14.3	-	-

Table 9:

Perimembranous VSD- physiological diagnosis						
	Right to right shunting		Left to right shunting		Bi-directional shunting	
	No.	%	No.	%	No.	%
Total sample (n=44)	15	34.1	9	20.4	15	34.1
< 2 year (n=11)	6	54.5	1	9.1	4	36.3
2-4 years (n=21)	5	23.8	6	28.6	8	38.1
> 4 years (n=12)	4	33.3	2	16.7	3	25

Table 10:

Outlet VSD- physiological diagnosis			
	Right to right shunting	Left to right shunting	Bi-directional shunting

	No.	%	No.	%	No.	%
Total sample (n=6)	3	50	2	33.3	1	16.7
< 2 year (n=4)	2	50	1	25	1	25
2-4 years (n=0)	-	-	-	-	-	-
> 4 years (n=2)	1	50	1	50	-	-

Table 11: Other cardiac abnormalities: < 2 years age group n=15 (30%)

Other cardiac abnormalities						
No.	Age	Sex	Race	General	Atrium	Ventricle
1.	0.67	M	Black	Cardiomegaly with apex beat in the 5th i.c.s. The 4 chambers of the heart were mildly enlarged. Mild infundibular stenosis	-	Double chamber RV. Muscle bundle present in the right ventricular cavity with turbulence across the band.
2.	0.75	F	Black	Cardiomegaly with the apex beat in the 6th i.c.s. Tight infundibular & PV stenosis, Mildly hypoplastic pulmonary annulus, Conal branch of RCA crossing RVOT. PFO	-	RV hypertrophied with a marked septal muscle bundle.
3.	0.83	M	Black	Small heart with apex beat in the 5th i.c.s. PDA. Dominant RCA. Branch of LCA crosses near RVOT.	Interatrial septum bulges towards the left atrium.	Tight os infundibuli. RVOT obstruction =fibro muscular tissue & muscle bands. Valve w as an outflow chamber. In posterior RV 2 muscle bands=double chamber RV.
4.	1.16	F	Black	PFO, Infundibular stenosis + pulmonary valve + suprapulmonary stenosis, Right sided aortic arch, Ductal bump on LPA	-	-
5.	1.16	M	Black	Cardiomegaly with apex beat in the 5th i.c.s. lateral MCL. Aortic + infundibular pulmonary stenosis, Small pulmonary arteries, Right sided artic arch. . Small 2mm PFO.	-	Hypertrophied LV + RV: dilated, hypertrophied & poorly contractile. Thick hypertrophied muscle bundles situated lower in RV than usually in TOF.
6.	1.17	F	Black	Hypoplastic MPA, Stenosis of proximal LPA, Tight infundibular stenosis, Stenotic pulmonary valve, Large fossa ovalis with PFO	4mm ASD	-
7.	1.33	F	Black	3mm PFO. Pulmonary valvar stenosis, severe infundibular narrowing	-	RV severely hypertrophied & there were markedly hypertrophied muscle bundles obstructing the infundibular area. Conal branch of RCA crossing RVOT
8.	1.42	F	Indian	3-4mm PFO. PDA=1-2mm. Infundibular & valvar pulmonary stenosis	-	RV pressure supra systemic suggesting a restrictive VSD. Narrowed RVOT & doming pulmonary valve. Right coronary artery crossing RVOT.

9.	1.5	F	Black	Small PFO.	RA dilated with inter atrial septum bulging to the left.	Large hypertrophied parietal & septal muscle bands. One of the aortic cusps seems to be attached to one of the septal bands.
10.	1.5	M	White	PFO. Subvalvar, valvar & supravalvar PS, Left sided aortic arch	-	Hypertrophied parietal septal muscle bundles
11.	1.5	F	Black	Mild infundibular stenosis. The 4 chambers of the heart were enlarged. Bilateral Harrison's sulci. Cardiomegaly with apex beat displaced to 6th l.i.c.s. lateral to MCL.	-	Large parietal muscle bundle present. Large papillary muscle was coming high of the septum as well. Biventricular hypertrophy.
12.	1.5	F	White	Subvalvar & valvar pulmonary stenosis, Right sided aortic arch	-	Large septal & parietal muscle bundles.
13.	1.58	M	Colored	Subvalvar & valvar pulmonary stenosis, Small PDA, Right sided aortic arch	-	Thick muscular infundibulum, Aberrant coronary on left crossing RVOT above the level of the PV+ another vessel that appears to cross RVOT at the level of the RVOT.
14.	1.58	M	Black	Small PFO. Boot shaped heart.	-	Small infundibular chamber & a tight infundibular os (4mm) surrounded by fibrous edges. Hypertrophied parietal & septal muscle bundles.
15.	1.58	F	Black	Hypoplastic MPA with severe pulmonary valve hypoplasia. PFO.	Small ASD with R to L shunt=5.8mm	Very high suprasystemic pressures in RV with high RVEDP indicating restrictive VSD. Large conal branch originating from RCA that goes towards the RVOT but not crossing it.

Table 12: Other cardiac abnormalities: 2-4 year age group n=21 (42%)

Other cardiac abnormalities						
No.	Age	Sex	Race	General	Atrium	Ventricles
1.	2	M	Black	-	-	RV hypertrophied with massive septal & parietal muscle bands.
2.	2	F	Colored	Infundibular stenosis Acyanotic tetralogy Right aortic arch, DSAS membrane of 3.6mm. Cardiomegaly with apex beat in the 5th i.c.s. lateral to the MCL.	-	Conal branch of RCA crosses RVOT, Moderate infundibular stenosis with hypertrophied parietal & septal muscle bundles
3.	2	F	Black	Small PFO. Hypoplastic pulmonary valve & MPA Origin RPA stenosis.	-	Small conal branches were crossing the RVOT.
4.	2	F	Colored	Infundibular stenosis, PFO present. Valvar & supravalvar stenosis. Cardiomegaly with the apex beat in the 5th l.i.c.s	Interatrial septum intact but bulges to the left	RV is tripartite and is dilated. Conal branch of the RCA crossing the RVOT.
5.	2	M	Black	Boot shaped heart. Tight infundibular stenosis	-	Small infundibular chamber with thick fibro muscular tissue
6.	2	F	Black	Mild infundibular stenosis, Apex beat in 5th ICS	-	Double chamber RV, There is a muscle band dividing the RV.

7.	2	M	Black	PFO=5-6mm. Cardiomegaly. Tight infundibular stenosis	-	LAD came of RCA and was seen crossing RVOT. RV hypertrophied with thick hypertrophied septal and parietal muscle bundles.
8.	2	M	Black	Infundibular, valvar & supravalvar stenosis, Dilated ascending aorta. PFO	Bulging to the left	RV hypertrophied with very thick parietal & septal muscle bundles. Small infundibular chamber with a tight os 2-3mm. Os had fibrous edges & small vegetations on os. Conal branch of RCC crossing the RVOT
9.	2	M	Black	Mild cardiomegaly. Infundibular stenosis	RA + LA enlarged	Infundibular stenosis. Large, thick muscle band from the base of the TV towards the free wall of RV obstructing RVOT Muscle bands dividing the RVOT into 2 narrow channels. Double chamber RV
10.	2	F	Black	Infundibular + valvular pulmonary stenosis, Right sided aortic arch, Innominate vein inferior to aortic arch. 4 chambers of the heart were dilated. . Apex beat in 5th ICS. Cardiomegaly	-	Hypertrophied parietal septal muscle bundles delineating an infundibular os (5mm)
11.	3	F	Black	Valvar pulmonary stenosis, PFO	-	Marked hypertrophied parietal & septal muscle bundles
12.	3	F	Black	Cardiomegaly with the apex beat in the 4th I.c.s. Small DSAS	-	Double chamber RV.
13.	3	M	Black	Infundibular & valvar pulmonary stenosis, Long tubular stenosis of the origin of LPA, Aortic override, Heart was mildly dextro rotated with the right SVC located posteriorly. PDA=1-2mm	-	Minimal hypertrophied right ventricular muscle bundles but RV was hypertrophied
14.	3	M	Black	Mild cardiomegaly, Infundibular Pulmonary stenosis	Right atrial enlargement	Tight infundibular stenosis with hypertrophied septal & parietal muscle bundles, Abnormal conal branching crossing RVOT
15.	3	M	Black	Tight infundibular pulmonary stenosis, Heart was markedly dilated rotated towards the right (mesocardia). Marked 4 chamber enlargement. Cardiomegaly	-	Large hypertrophied parietal & septal muscle bands leaving several channels & making the true infundibular os difficult to find.
16.	3	M	Black	Right sided aortic arch, DSAS, 4 chambers of the heart were dilated. Cardiomegaly with apex in the 5th l.i.c.s.	-	Biventricular hypertrophy. Double chambered RV with opening=9mm.
17.	3	M	Black	PDA=2.5mm. Sinus venosus, Pulmonary + infundibular stenosis, Dysplastic pulmonary valve	RA enlarged, ASD=6mm. Bi-directional shunting	-

18.	4	M	Black	Moderate to severe pulmonary stenosis & regurgitation Right sided aortic arch, Pulmonary valvar stenosis, Dense adhesions between the sternum & the heart. Boot shaped heart with a pulmonary bay, PFO=5mm.	Interatrial septum bulges to the left	Branch of RCA crossing RVOT
19.	4	M	Black	Mild cardiomegaly with apex displaced to 5th l.i.c.s, The 4 chambers of the heart were dilated	-	Double chamber RV
20.	4	M	White	-	-	Coronary crossing RVOT, Severe infundibular stenosis. Double outlet RV. Large conal branch descends on the infundibulum.
21.	4	F	Black	Cardiomegaly with the apex beat in the 5th l.i.c.s laterally displaced. Tight infundibular stenosis	-	Tiny conal branch of the RCA crossing the RVOT.

Table 13: **Other cardiac abnormalities: > 4 years age group n=14 (28%)**

Other cardiac abnormalities						
No.	Age	Sex	Race	General	Atrium	Ventricles
1.	5	F	Black	Infundibular stenosis, Valvar stenosis, PFO, PDA=1-2mm	Large ASD secundum =6-7mm (op).	-
2.	5	F	Black	PFO. Pulmonary valvar stenosis	-	RV globally hypertrophied. No marked infundibular hypertrophied muscle bundles
3.	5	F	Colored	Absent LPA, Hypoplastic MPA, Right sided aortic arch, The innominate vein was not in the usual place but was found to run under the aortic arch behind the ascending aorta & joined the SVC anteriorly & superiorly to the RPA. PFO=4-5mm	1mm perforation seen in the atrial septum along the medial rim	Large coronary branch seen crossing RVOT.
4.	5	M	Black	PFO=2mm. Hypoplastic pulmonary valve + MPA	-	-
5.	5	M	Colored	Infundibular stenosis, Possible MPA hypoplasia	-	-
6.	5	M	Black	Subvalvar + valvar + supravalvar pulmonary stenosis, Hypoplastic LPA, Right sided heart failure	-	-
7.	6	M	Black	The 4 chambers of the heart were dilated. Moderate aortic regurgitation, DSAS	-	Double chamber RV
8.	6	M	Black	Discrete subaortic membrane, Severe infundibular stenosis, PFO=1-2mm.	-	Extremely hypertrophied septal & parietal muscle bundles (papillary muscle & chords attached to 1). 2-3mm infundibular os with small fibrous vegetations on edge.
9.	6	M	Black	-	-	RCA crossing RVOT
10.	6	M	Black	Absent LPA, Valvar & supravalvar stenosis. Cardiomegaly with the apex beat in the 5th I.c.s. lateral to the MCL	-	RV was dilated & hypertrophied. No infundibular hypertrophic muscle bundles.

11.	7	M	Black	Tight infundibular stenosis, Anomalous right subclavian artery arising from the descending aorta, Apex beat in the 5th I.c.s. Small PFO = 2-3mm.	Interatrial septum bulges to the left.	Biventricular enlargement
12.	9	F	Black	Right sided SVC and innominate vein. Rotated to the left with left ventricle being very posterior and difficult to see. Tight infundibular stenosis, Very small pulmonary annulus.	-	Small conal branch of the RCA crossing RVOT. Small echo densities within the RV, which have the appearance of tiny vegetations. RV very hypertrophied
13.	9	F	Black	Pulmonary atresia, Infundibular & valvar PS, Right sided aortic arch, Absent LPA. Cardiomegaly with apex beat in 5th l.i.c.s.	-	RV small & moderately hypertrophied.
14.	13	F	Black	PV+ infundibular stenosis, Ductal bump on LPA, Sterile vegetations within infundibulum, Mesocardia	-	Severe infundibular stenosis due to a thick anterior wall. Possible infundibular vegetations

Table 14: Other thoracic abnormalities: < 2 years age group n=15 (30%)

No.	Age	Sex	Race	Other thoracic abnormalities
1.	0.67	M	Black	Pulmonary hypertension, LPSH. Hyperinflated lungs.
2.	0.75	F	Black	LPSH & a precordial bulge. Minor Harrison's sulci bilaterally
3.	0.83	M	Black	Oligaemic lung fields
4.	1.16	F	Black	URTI with rhinorrhoea. Mild Harrison's sulcus & mild pectus carrinatum
5.	1.16	M	Black	Mild pectus carrinatum. Left lateral thoracotomy scar.
6.	1.17	F	Black	Oligaemic lung fields.
7.	1.33	F	Black	Oligaemic lung fields
8.	1.42	F	Indian	-
9.	1.5	F	Black	Rotated chest. Oligaemic lung fields.
10.	1.5	M	White	Oligaemic lung fields
11.	1.5	F	Black	Mild pulmonary hypertension, Pectus carrinatum, LPSH. Plethoric lung fields
12.	1.5	F	White	LPSH. Oligaemic lung fields.
13.	1.58	M	Colored	-
14.	1.58	M	Black	Mild Harrison's sulcus. LPSH + epigastric pulsation indicating RVH. Oligaemic lung fields.
15.	1.58	F	Black	-

Table 15: Other thoracic abnormalities: 2-4 year age group n=21 (42%)

No.	Age	Sex	Race	Other thoracic abnormalities
1.	2	M	Black	-
2.	2	F	Colored	-
3.	2	F	Black	-
4.	2	F	Colored	Bilateral Harrison's sulcus
5.	2	M	Black	-
6.	2	F	Black	Mild pulmonary hypertension, left parasternal heave.
7.	2	M	Black	Oligaemic lungs
8.	2	M	Black	-
9.	2	M	Black	Plethoric lung fields
10.	2	F	Black	Plethoric with recurrent infective changes. Precordial bulging
11.	3	F	Black	-
12.	3	F	Black	-
13.	3	M	Black	-

14.	3	M	Black	-
15.	3	M	Black	Oligaemic lung fields.
16.	3	M	Black	Bilateral Harrison's sulcus.
17.	3	M	Black	-
18.	4	M	Black	-
19.	4	M	Black	Left parasternal heave + epigastric pulsation indicating RVH.
20.	4	M	White	-
21.	4	F	Black	Left parasternal heave. Lung fields are Oligaemic

Table 16: Other thoracic abnormalities: > 4 years age group n=14 (28%)

No.	Age	Sex	Race	Other thoracic abnormalities
1.	5	F	Black	-
2.	5	F	Black	-
3.	5	F	Colored	Plethoric lung fields.
4.	5	M	Black	Plethoric lung fields. LPSH.
5.	5	M	Colored	-
6.	5	M	Black	Pulmonary hypertension
7.	6	M	Black	-
8.	6	M	Black	Mild Harrison's sulcus. Oligaemic lung fields, murmur radiates to the lungs.
9.	6	M	Black	Hyperinflated chest with Harrison's sulcus. Mild respiratory distress with kussmaul breathing. Oligaemic lung fields. Obvious precordial bulge. Left parasternal heave
10.	6	M	Black	LPSH.
11.	7	M	Black	-
12.	9	F	Black	Bilateral Harrison's sulcus. Oligaemic lung fields.
13.	9	F	Black	Moderate pulmonary hypertension in right lung, Plethoric lung fields. Left precordial bulge. Left parasternal heave + epigastric pulsation indicating RVH. Hypoplastic left lung
14.	13	F	Black	Severe pulmonary hypertension, Mild LPSH

Table 17:

Other diagnoses: < 2 years age group n=15 (30%)					
No.	Age	Sex	Race	2.4 Genetic abnormalities & syndromes	2.5 Other
1.	0.67	M	Black	-	-
2.	0.75	F	Black	-	Clubbing of all digits. Mildly cyanosed Hypercyanotic spells,
3.	0.83	M	Black	-	Anemia, Severe hypercyanotic spells
4.	1.16	F	Black	-	Mild dysmorphic features with a flat nasal bridge & epicanthic folds & hypertelorism. Fe deficiency anemia. Hypecyanotic spells
5.	1.16	M	Black	-	3cm pulsatile liver: 2 left lobes-left upper & left lower. Soft 1cm hepatomegaly. No left SVC. Digital clubbing. Subtle dysmorphism-low set simple ears. Severe cyanotic spells
6.	1.17	F	Black	-	Shotty inguinal lymphadenopathy. Clubbing of all digits. No innominate vein seen. Hypercyanotic spells
7.	1.33	F	Black	-	Severe polycythemia, Thrombocytopenia- hypercyanotic spells
8.	1.42	F	Indian	-	-
9.	1.5	F	Black	-	Hypercyanotic spells Clubbing of the digits. Crying incessantly
10.	1.5	M	White	-	Delayed milestones: unable to sit unsupported & not yet crawling. Not thriving well.
11.	1.5	F	Black	-	Large conal branch coming of the LAD and dividing further in 3 significant branches

12.	1.5	F	White	-	Dysmorphic looking with a round face. Wide epicanthic folds & slightly deep set eyes. Clubbing
13.	1.58	M	Colored	-	Hypercyanotic spells, Subtle dysmorphic features
14.	1.58	M	Black	-	Digital clubbing. Epicanthic folds but no gross dysmorphic features. Hypercyanotic spells
15.	1.58	F	Black	-	Severe hypocyanotic spell. Severe cyanosis & clubbing of digits

Table 18:

Other diagnoses: 2-4 year age group n=21 (42%)					
No.	Age	Sex	Race	2.4 Genetic abnormalities & syndromes	2.5 Other
1.	2	M	Black	-	Cyanotic spells, Ambiguous genitalia Undervirilised male, Large conal branch coming from RCA. Dry gangrene of the toes & distal parts of both feet. Sternal sepsis. Small genitalia & undescended testis.
2.	2	F	Colored	-	-
3.	2	F	Black	-	-
4.	2	F	Colored	-	Hypercyanotic spells
5.	2	M	Black	-	Early digital clubbing. Acyanotic Hypercyanotic spells
6.	2	F	Black	-	Left kidney is ectopic situated in the pelvic region.
7.	2	M	Black	-	Slightly dysmorphic. Clubbed, Hypercyanotic spells
8.	2	M	Black	-	Hypercyanotic spells. Digital clubbing. Soft hepatomegaly 2cm
9.	2	M	Black	-	-
10.	2	F	Black	-	Mild digital clubbing. Anomalous innominate vein (a large vein was coursing under the aortic arch just anteriorly and superiorly to RPA and was draining into left side above RA/SVC junction.
11.	3	F	Black	-	Episodic cyanotic spells, Mildly cyanosed & mild clubbing.
12.	3	F	Black	-	-
13.	3	M	Black	-	-
14.	3	M	Black	-	Acyanotic
15.	3	M	Black	-	Hypercyanotic spells, Subtle dysmorphic features-epicanthic folds. Digital clubbing. Plethoric conjunctivae.
16.	3	M	Black	-	-
17.	3	M	Black	-	Anomalous azygos vein
18.	4	M	Black	-	Sternotomy & bilateral thoracotomy scar. Severe cyanosis & clubbing of the digits. Hoarse voice with hoarse cough
19.	4	M	Black	-	-
20.	4	M	White	-	-
21.	4	F	Black	-	Digital clubbing. Hypercyanotic spells

Table 19:

Other diagnoses: > 4 years age group n=14 (28%)					
No.	Age	Sex	Race	2.4 Genetic abnormalities & syndromes	2.5 Other
1.	5	F	Black	-	-
2.	5	F	Black	-	Acyanotic
3.	5	F	Colored	-	Hypercyanotic spells, Clubbing.
4.	5	M	Black	-	3cm hepatomegaly. Acyanotic
5.	5	M	Colored	-	Acyanotic

6.	5	M	Black	Mendelssohn syndrome (aspiration of gastric contents)	Renal dysfunction, Severe oedema (anarsarca)
7.	6	M	Black	-	-
8.	6	M	Black	-	Severe cyanotic spells, Height & weight below percentile
9.	6	M	Black	-	Digital clubbing of fingers & toes. Z score= -3. Hypercyanotic spells, Mild convergent strabismus, Speech & hearing problems.
10.	6	M	Black	-	Hypercyanotic spells
11.	7	M	Black	Catch 22 positive	Old left hemiplegia, Left clubbed foot, walks with a hemiplegic gait. Subtle dysmorphic features. Mild cyanosis.
12.	9	F	Black	-	Gross clubbing of the digits, Polycythemia
13.	9	F	Black	-	Right thoracotomy scar. Mild digital clubbing.
14.	13	F	Black	-	Clubbing of all digits. Cyanosis. Tanner stage 2 breast development

First presentation of the VSD and the operation details

Table 20: First presentation of the VSD: < 2 years age group n=15 (30%)

No.	Age	Sex	Race	Reason for initial VSD diagnoses	Reason for delaying repair	Time lapse between first presentation& op
1.	0.67	M	Black	Child was referred to clinic for work up of incidental murmur. Assessed to have PM VSD with bi-directional shunting across defect.	Waiting list	8 months
2.	0.75	F	Black	Diagnosed early to have VSD with TOF.	Waiting list	7 months
3.	0.83	M	Black	Presented with gastroenteritis & broncho pneumonia. Noted to have cyanotic episodes	Waiting list	1 month
4.	1.16	F	Black	The child was seen for episodes of shortness of breath, which may have been cyanotic spells.	Waiting list	1 month
5.	1.16	M	Black	Referred with history of severe respiratory distress.	Urgent shunt requested & operation done as life saving procedure (child too critical to undergo cardiac cath to identify anatomy) to reduce cyanosis before VSD could be repaired.	2 months
6.	1.17	F	Black	Diagnosed as TOF at birth	Waiting list	15 months
7.	1.33	F	Black	First presented with neonatal sepsis & murmur.	Waiting list	17 months
8.	1.42	F	Indian	Referred for assessment of cardiac murmur. Blue on exertion. Assessed to have TOF like anatomy.	Waiting list	4 months
9.	1.5	F	Black	Admitted with severe cyanotic spells & abdominal cramps. Typical TOF anatomy.	Waiting list	2 months
10.	1.5	M	White	Referred with incidental murmur.	Waiting list	6 months
11.	1.5	F	Black	Assessed to have cardiac murmur & found to have TOF anatomy.	Waiting list	4 months
12.	1.5	F	White	Noted to be cyanosed with episodes of squatting. Diagnosed to have TOF anatomy.	Waiting list	2 month
13.	1.58	M	Colored	History of productive cough since birth. Hypercyanotic spells	Waiting list	9 months
14.	1.58	M	Black	Referred for assessment of cardiac murmur. Admitted for meningoencephalitis found to have incidental cardiac murmur	Waiting list	2 months
15.	1.58	F	Black	Referred for repair of defect, also had acute gastroenteritis.	Waiting list	9 months

Table 21: First presentation of the VSD: 2-4 year age group n=21 (42%)

No.	Age	Sex	Race	Reason for initial VSD diagnoses	Reason for delaying repair	Time lapse between first presentation& op
1.	2	M	Black	Assessed to have an acyanotic TOF but has become cyanosed.	Waiting list	17 months
2.	2	F	Colored	Referred to cardiac clinic with an incidental murmur.	Waiting list	22 months
3.	2	F	Black	Mother noticed child to be cyanosed when she was 6 months old. Investigation revealed an atypical TOF in that there was no infundibular stenosis but stenosis at valvar & supravalvar level. Also RPA stenosis.	Waiting list	5 month
4.	2	F	Colored	Diagnosed as TOF in infancy. History of cyanotic spells.	Waiting list	2 years & 9 months
5.	2	M	Black	Hypercyanotic spells	Waiting list	1 month
6.	2	F	Black	Initially presented with URTI & an incidental murmur was picked up. Thought to have TOF like anatomy.	Waiting list	8 months
7.	2	M	Black	Cyanosed with frequent cyanotic spells	Waiting list	3 months
8.	2	M	Black	Child seen shortly after birth because of respiratory distress. Diagnosis of VSD with mild pulmonary stenosis made. History of cyanotic spells	Waiting list	4 months
9.	2	M	Black	Followed up since 7 months old, diagnosed as non- cyanotic TOF.	Waiting list	3 months
10.	2	F	Black	Brought in for assessment of cardiac murmur when child had an URTI.	Waiting list	1 month
11.	3	F	Black	Child initially presented as an acyanotic TOF.	Waiting list	2 years & 9 months
12.	3	F	Black	Referred for incidental finding of a cardiac murmur.	Waiting list	20 months
13.	3	M	Black	Presented with bronchopneumonia & sent for assessment of cardiac murmur.	Waiting list	2 years & 10 months
14.	3	M	Black	Referred by GP for assessment of cardiac murmur. Assessed to have TOF anatomy	Waiting list	7 months
15.	3	M	Black	Child presented with history of cough, dyspnea & worsening cyanosis	Waiting list	1 month
16.	3	M	Black	Referred for evaluation of incidental murmur. Recurrent URTI`s	Waiting list	1 month
17.	3	M	Black	First seen at age 7 weeks when was referred for assessment of cardiac murmur suspected to be VSD.	Waiting list	14 months
18.	4	M	Black	Assessed to have TOF with hypoplastic MPA & hypercyanotic spells.	RVOT inserted first then waiting list for VSD correction	4 years & 9 months
19.	4	M	Black	Referred for acute gastritis & cardiac murmur was heard. Assessed to have PM VSD & DCRV	Waiting list	2 years
20.	4	M	White	Severe cyanosis and spells.	BT shunt more urgent than repair of VSD.	2 years & 4 months
21.	4	F	Black	Referred for cyanosis & incidental murmur. History of effort intolerance & frequent episodes	Waiting list	12 months

				of squatting.		

Table 22: First presentation of the VSD: > 4 years age group n=14 (28%)

No.	Age	Sex	Race	Reason for initial VSD diagnoses	Reason for delaying repair	Time lapse between first presentation& op
1.	5	F	Black	Assessed to have acyanotic TOF. Also had infundibular & valvar pulmonary stenosis.	Waiting list	3 years & 1 month
2.	5	F	Black	Presented with an acyanotic TOF. Typical TOF anatomy with infundibular stenosis & thickened doming PV & good pulmonary arterial branches. Treated for SBE.	Waiting list	2 years & 1 month
3.	5	F	Colored	Assessed to have complex TOF.	Was admitted for surgery but mother was afraid & took child home	3 years & 2 months
4.	5	M	Black	Known to have cardiac murmur since birth. History of chronic bronchitis. Assessed to have a TOF.	Waiting list	1 month
5.	5	M	Colored	Referred for incidental murmur.	Waiting list	8 months
6.	5	M	Black	Downs syndrome, found to have a non- restrictive VSD & small PDA.	Waiting list	6 months
7.	6	M	Black	Admitted with bronchopneumonia & CCF. Failed to thrive. Assessed to have large mid muscular VSD measuring 19.7mm on LV side & narrows down to 8.9mm on RV side.	Waiting list	9 months
8.	6	M	Black	Presented with history of cyanosis & squatting.	Lost to follow up presented again now. Put on waiting list for semi urgent surgery.	2 years & 10 months
9.	6	M	Black	Diagnosed with TOF & was booked for surgery. Severe cyanotic spells, was pyrexial & SBE treatment as started.	Mother did not bring child in.	3 years & 3 months
10.	6	M	Black	Seen after fainting episode of 10 min duration, history of cough, fever, vomiting and haemoptysis. Mildly desaturated & assessed to have TOF with absent LPA. History of paroxysmal nocturnal dyspnea.	Waiting list	3 months
11.	7	M	Black	Initially referred for TOF & left hemiplegia. Cyanosed since infancy	Waiting list	3 years
12.	9	F	Black	Referred for assessment of cyanotic congenital heart problem. Diagnosed as TOF.	Waiting list	2 months
13.	9	F	Black	Presented to cardiac clinic with severe cyanosis. Assessed as pulmonary atresia & VSD then sent for urgent B-T shunt. In 1999 found to have TOF with absent LPA & hypertensive RPA.	BT shunt more urgent than repair of VSD.	6 years
14.	13	F	Black	Seen due to shortness of breath with exertion	Waiting list	5 months

Table 23: Operation details: < 2 years age group n=15 (30%)

No.	Age	Sex	Race	Type of procedure that was used to repair the VSD
1.	0.67	M	Black	Closure of VSD with patch of vascular goretex. Resection of septal muscle bundle.
2.	0.75	F	Black	Complete correction of TOF. Closure of VSD with patch of vascular goretex. Left a 4mm PFO.
3.	0.83	M	Black	Correction of TOF: Enlargement of MPA & RV outflow tract with monocusp valve patch. Creation of small ASD.VSD closed with goretex patch
4.	1.16	F	Black	Complete repair of TOF: Closure of VSD with vascular goretex patch. Widening of RVOT. Left a 4mm PFO.
5.	1.16	M	Black	Complete correction of TOF: Left thoracotomy to free left BT shunt. Closure of small PM VSD using vascular goretex patch. Resection of large RVOT muscle bundles. RVOT patch. Calibrated PFO 4mm.
6.	1.17	F	Black	VSD closed with Goretex patch. Enlargement of TVOT with bovine pericardium patch. Enlargement of MPA & LPA with bovine pericardium.
7.	1.33	F	Black	VSD closed with patch of bovine pericardium. Hypertrophied muscle bundles resected. RVOT widened with patch of bovine pericardium.
8.	1.42	F	Indian	Closure of true subarterial VSD with bovine pericardial patch. PDA ligation. Transannular RVOT patch. Left a 3-4mm PFO.
9.	1.5	F	Black	Complete correction of TOF: closure of VSD with vascular goretex patch. Patch widening of RVOT. PFO left open.
10.	1.5	M	White	Complete correction of TOF: VSD closed with patch of vascular goretex. Widening of RVOT + MPA.
11.	1.5	F	Black	Closure of VSD with vascular goretex patch. Excision of parietal muscle bundle. Widening of RVOT with vascular goretex patch.
12.	1.5	F	White	Closure of large malaligned PM VSD with Goretex patch. Widening RVOT with an aortic homograph monocusp. Left a PFO.
13.	1.58	M	Colored	Repair of TOF: Closure of PM VSD with goretex patch. Resection of muscle bundles, RVOT enlargement. PDA ligation.
14.	1.58	M	Black	Complete correction of TOF. Closure of VSD with patch of vascular goretex. Left PFO open.
15.	1.58	F	Black	Complete repair of TOF: Goretex patch closure of large muscular PM outlet VSD & calibrated 4mm ASD.

Table 24: Operation details: 2-4 year age group n=21 (42%)

No.	Age	Sex	Race	Type of procedure that was used to repair the VSD
1.	2	M	Black	Total correction of TOF: Closure of subarterial VSD with vascular goretex patch, resection of hypertrophied muscle bands. Transannular bovine pericardium. RVOT patch. Left a 3-4mm PFO.
2.	2	F	Colored	Complete correction of TOF: Closure of VSD using goretex patch & resection of sub aortic membrane
3.	2	F	Black	Total correction of TOF: Closure of VSD with vascular goretex patch Left a 4mm PFO
4.	2	F	Colored	Complete correction of TOF: closure of VSD with vascular goretex patch, widening of RVOT & MPA with transannular patch of bovine pericardium up to the bifurcation. Left 4mm calibrated PFO open.
5.	2	M	Black	Calibrated 4mm PFO. VSD closed with patch of vascular goretex.
6.	2	F	Black	PFO surgically created & LV entered via PFO & LA. TV retracted to expose VSD. Infundibulotomy then performed, thick parietal muscle bundle excised. VSD closed with prolene suture.
7.	2	M	Black	Complete correction of TOF: Closed of VSD with bovine pericardium patch, bovine pericardium ventriculotomy patch, and PFO left open.
8.	2	M	Black	Patch closure of VSD using vascular goretex, transannular patch widening of RVOT. Left a PFO.

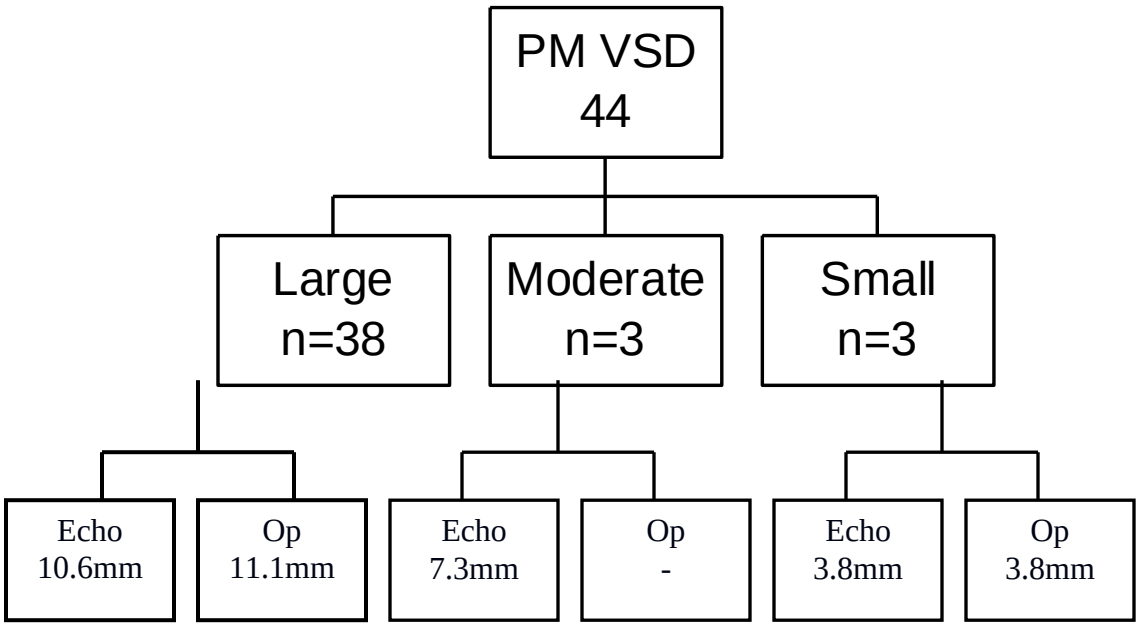
9.	2	M	Black	Closure of VSD with vascular goretex patch, Relief of RV outflow tract obstruction: excision of RV muscle band
10.	2	F	Black	Complete correction of TOF. VSD closed with patch of vascular goretex.
11.	3	F	Black	Complete correction of TOF. VSD closed with goretex vascular patch.
12.	3	F	Black	Patch closure of VSD using bovine pericardium. Resection of fibrous os causing DCRV.
13.	3	M	Black	Complete correction of TOF: Repair of origin of LPA stenosis with patch of pulmonary homograph tissue, widening of RVOT & MPA with monocusp pulmonary homograph. VSD closed with Goretex patch.
14.	3	M	Black	Complete correction of TOF: VSD closed with vascular goretex patch.
15.	3	M	Black	Complete correction of TOF: transannular RVOT patch. Creation of PFO. VSD closed with vascular goretex patch.
16.	3	M	Black	Closure of VSD with vascular goretex patch. Resection of RV parietal muscle bundle. Fibrous obstructing tissue in RVOT resected & hypertrophied muscle band divided.
17.	3	M	Black	Ligation of PDA, closure of VSD with patch of vascular goretex, enlargement of RVOT & PA with monocucp valve & bovine pericardial patch, primary closure of ASD
18.	4	M	Black	Complete correction of TOF: VSD closed with vascular goretex patch. Repair of RVOT.
19.	4	M	Black	Closure of VSD with patch of goretex. Resection of infundibular obstructive muscle bundles
20.	4	M	White	Ligation of central shunt. Closure of VSD with vascular goretex patch. Left 4mm PFO.
21.	4	F	Black	Closure of VSD through ventriculotomy with bovine pericardium. Infundibulotomy, insertion of 16mm homograph in RVOT, creation of 4mm PFO.

Table 25: Operation details: > 4 years age group n=14 (28%)

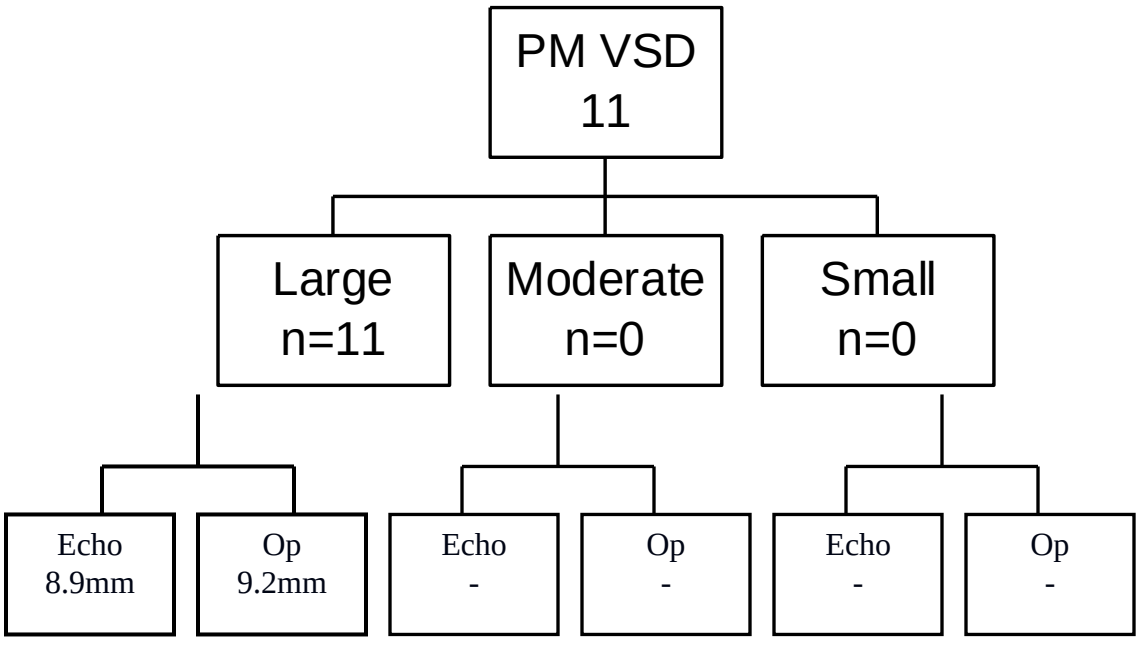
No.	Age	Sex	Race	Type of procedure that was used to repair the VSD
1.	5	F	Black	PDA ligation, closure of PM VSD with bovine pericardial patch, widening of the RVOT + pulmonary annulus + MPA with a monocusp from a size 20mm pulmonary homograph. Partial closure of secundum ASD leaving 3-4mm PFO.
2.	5	F	Black	Correction of TOF: closure of VSD with vascular goretex patch. Pulmonary valvotomy.
3.	5	F	Colored	Complete repair of TOF: closure of VSD with vascular goretex. Replacement of PV & MPA. PFO left open.
4.	5	M	Black	Complete correction of TOF: Closure of VSD with vascular goretex patch. Widening RVOT. Left a small PFO.
5.	5	M	Colored	Complete correction of TOF: closure of PM malaligned VSD with bovine pericardial patch, widening of RVOT with bovine pericardium up to bifurcation. Left 4mm calibrated PFO open.
6.	5	M	Black	Closure of residual VSD with vascular goretex patch. Insertion of ecmo oxygenator post op due to severe hypoxemia
7.	6	M	Black	Closure of subaortic VSD with vascular goretex patch. Resection of infundibular muscle bundles. Resection of DSAS. Aortic valve repair.
8.	6	M	Black	VSD closed with vascular goretex patch.
9.	6	M	Black	Complete correction of TOF: closure of malaligned PM VSD. Widening of RVOT.
10.	6	M	Black	Complete correction of TOF: closure of large malaligned PM VSD with vascular goretex. Widening of RVOT & MPA. Left a 4mm PFO.
11.	7	M	Black	Closure of true subarterial VSD with bovine pericardial patch. Widening of RVOT + pulmonary annulus + MPA with a trans annular patch of bovine pericardium. Left a 4mm PFO.
12.	9	F	Black	Complete correction of TOF: Resection of infundibular muscle bundles. Closure of VSD with patch of vascular goretex. PV replacement with 21mm Hancock bioprosthesis & widening of RVOT & MPA.
13.	9	F	Black	Ligation of BT shunt. Complete correction of TOF. VSD closed with vascular goretex patch.
14.	13	F	Black	Repair of TOF: PM VSD closed with patch of vascular goretex. Pulmonary valve enlargement.

VSDs at operation

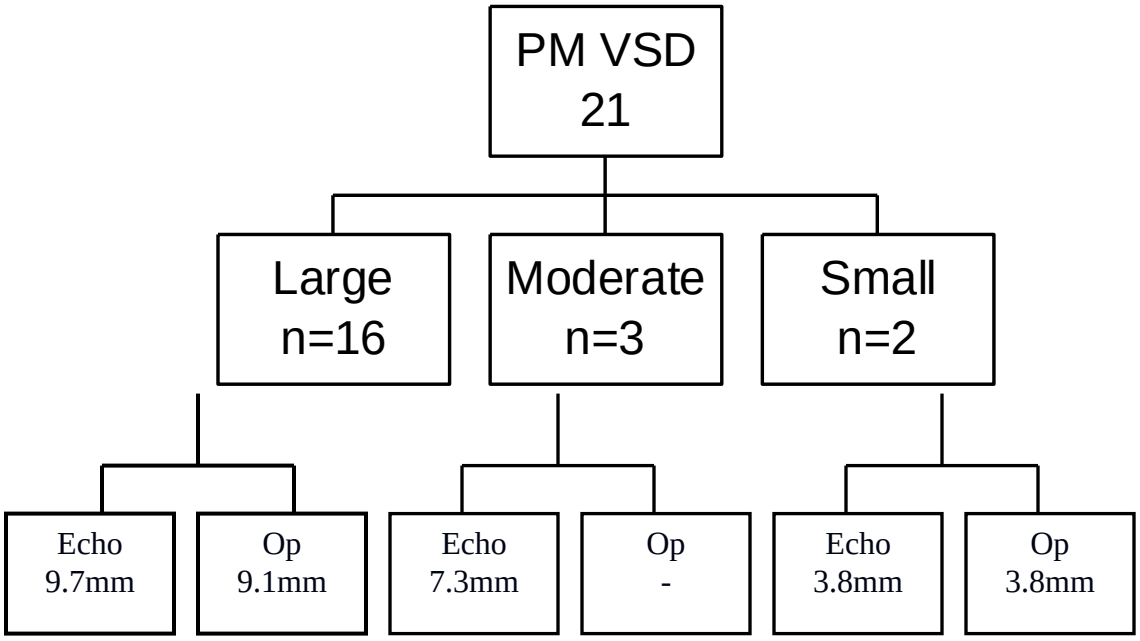
Types & sizes of the VSDs: Total sample n=50 (100%)



Types & sizes of the VSDs: < 2 years age group n=15 (30%)



Types & sizes of the VSDs: 2-4 year age group n=21 (42%)



Types & sizes of the VSDs: > 4 years age group n=13 (26%)

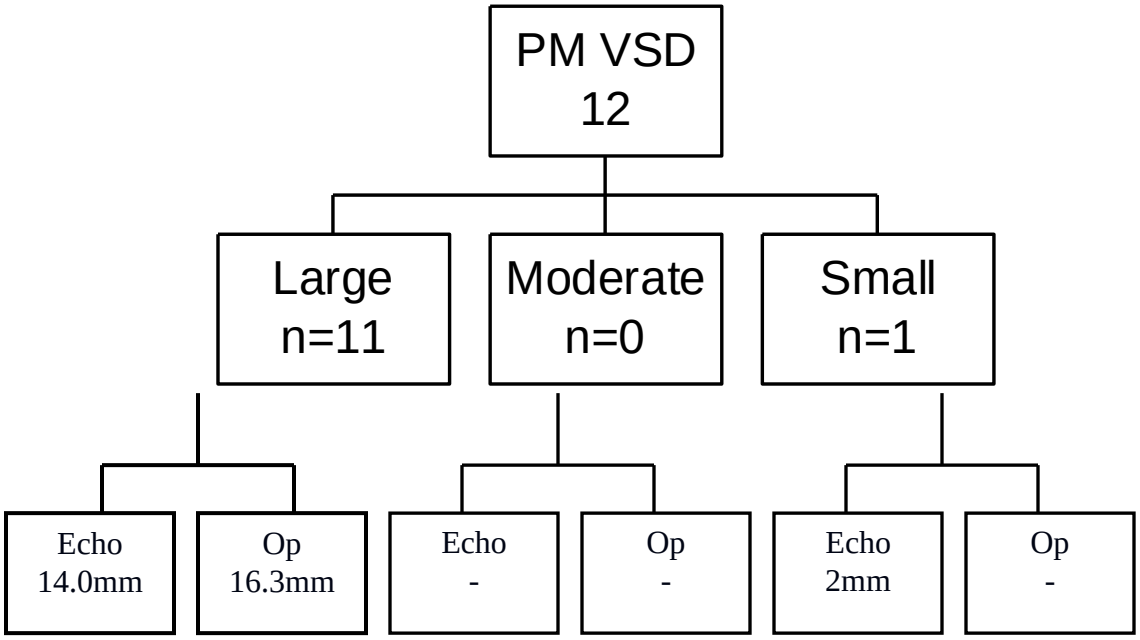


Table 26: Types & sizes of the VSDs

Outlet VSDs				
	Total sample N= 6	< 2 years age group n=4	2-4 year age group n=0	> 4 years age group n=2
Echo (mm)	10.2	7.3	-	14.5
Operation (mm)	-	-	-	-

Location & detailed description of the VSD in the inter ventricular septum: Perimembranous VSDs n =44

Table 27: Large PM VSD n=38

No	Age	Sex	Race	**Location & description of VSD- Large Perimembranous**
1.	0.67	M	Black	Large PM VSD.
2.	0.75	F	Black	Large non restrictive VSD
3.	1.16	F	Black	Large non restrictive VSD
4.	1.16	M	Black	Large PM VSD with fibrous inferior border
5.	1.17	F	Black	Large PM VSD
6.	1.33	F	Black	PM malaligned VSD
7.	1.5	F	Black	PM VSD in subaortic position separated from TV by bar of muscle
8.	1.5	M	White	Large non-restrictive PM malaligned VSD. Antero septal tricuspid commissural chords crossing VSD.
9.	1.5	F	White	Malaligned PM VSD
10.	1.58	M	Colored	Large PM VSD with large overriding aorta
11.	1.58	M	Black	Large non-restrictive PM malaligned VSD. Antero- septal commissural chords of TV crossing VSD.
12.	2	F	Black	VSD large & non –restrictive, ventricular pressures were balanced.
13.	2	F	Colored	Large non- restrictive malaligned PM VSD. Small PFO present.
14.	2	M	Black	Large PM VSD with outlet extension
15.	2	M	Black	Atypical VSD in PM area with outlet extension, bar of muscle present between TV & VSD. Also separated from infundibular area by hypertrophied muscle bundles.
16.	2	M	Black	-
17.	2	M	Black	VSD seen below superior leaflets of TV. Chordae from leaflet partially obstructing VSD which was somewhere towards AV & had rim of muscle between it & PM septum. VSD had fibrous edges.
18.	2	F	Black	VSD large, malaligned, in PM position with outlet extension.
19.	3	F	Black	Large PM malaligned VSD
20.	3	M	Black	Large malaligned PM VSD. Hypertrophied muscle bundles in RVOT.
21.	3	M	Black	VSD, large & malaligned. Bar of muscle present between TV & VSD.
22.	3	M	Black	Septal tricuspid chord crossing VSD. Large hypertrophied parietal muscle bundle in outflow area of RV forming orifice of 6mm, surrounded by thick fibrous tissue some adherent to AV annulus. Small fibrous membrane seen along inferior edge of VSD on LV side.
23.	3	M	Black	VSD in PM area with subarterial extension
24.	4	M	Black	Large PM malaligned VSD
25.	4	M	Black	PM VSD in proximal chamber of RV, surrounded by fibrous edges. TV partially closing VSD, chords crossing VSD. Thick infundibular muscle bundles mainly thick septal muscle bundles & smaller parietal muscle bundles.
26.	4	M	White	VSD remote from PA, committed to AV but a few centimeters remote from it.
27.	4	F	Black	VSD PM in nature with outlet extension. Not much muscle bundle to incise or excise in the infundibulum.
28.	5	F	Black	Hypertrophied septal & parietal muscle bundles in RVOT, did not seem to form very tight stenosis. VSD typical large PM malaligned VSD.

29.	5	F	Black	Large PM VSD
30.	5	F	Colored	Large malaligned PM VSD
31.	5	M	Colored	Large PM VSD. Some hypertrophied muscle bundles in RVOT.
32.	5	M	Black	Large PM VSD situated in outlet/PM area
33.	6	M	Black	Large malaligned PM VSD. Small ridge of fibrous tissue present along infero lateral border of VSD on LV side.
34.	6	M	Black	Large non-restrictive VSD. Hyper echoic mass seen in subvalvar area, vegetations?
35.	6	M	Black	Massive malaligned PM VSD
36.	9	F	Black	Large PM VSD. VSD separated from TV by thick bar of muscle. Fibrous membrane present on inferior edge of VSD. Commissural chords of TV crossing VSD, superiorly small muscle bundle coming from subaortic conus crossed VSD.
37.	9	F	Black	Large PM VSD.
38.	13	F	Black	Malaligned PM VSD

Table 28: Moderate PM VSD n= 3

No	Age	Sex	Race	**Location & description of VSD- Moderate Perimembranous**
1.	2	M	Black	VSD in subarterial position.
2.	2	F	Colored	VSD large malaligned. Right anterior cusp of AV prolapses into VSD. Small fibrous membrane seen on infero- posterior edge of VSD on LV side.
3.	3	M	Black	PM VSD

Table 29: Small PM VSD n= 3

No	Age	Sex	Race	**Location & description of VSD- Small Perimembranous**
1.	2	F	Black	VSD in PM area & very small. Surrounded by thick fibrous tissue. Septal leaflet of TV in area of antero- septal commissure was thickened. There was a thick parietal muscle bundle in the infundibular area leaving an os of about 4-5mm with some fibrous edges in the higher chamber.
2.	3	F	Black	VSD in PM area & malaligned. Attempted closure of VSD by tricuspid leaflet. Superior border of VSD formed by aortic leaflets. Fibrous rim around VSD.
3.	5	M	Black	VSD small, malaligned & in PM position.

Table 30: Location & detailed description of the VSD in the inter ventricular septum: Outlet VSDs n=6

No.	Age	Sex	Race	**Location & description of VSD - outlet**
1.	0.83	M	Black	Large outlet VSD
2.	1.42	F	Indian	True subarterial VSD with conjoint aortic & pulmonary annuli. Hypertrophied muscle bundles mainly on septal side & few on parietal side.
3.	1.5	F	Black	True subarterial outlet VSD with conjoint aortic & pulmonary annuli. Anterior aortic valve cusp prolapsing into VSD practically occluding it.
4.	1.58	F	Black	Underdeveloped conal septum with outlet VSD. Right anterior cusp of aorta prolapsing into defect.
5.	6	M	Black	VSD subaortic with superior edge common to aortic annulus. RCC prolapsing into VSD & thickened with elongated free edge especially towards left cusp. Conus septum present with PV being distant from VSD & aortic annulus. Thick infundibular muscle bundle present with fibrous os. Thick subaortic membrane present under AV partially adherent to LCC.
6.	7	M	Black	True subarterial VSD. Hypertrophied parietal & septal muscle bundles in RVOT. Bar of muscle between inferior edges of VSD & TV. No infundibular muscle between upper edges of VSD & PV.

Appearance of the VSDs when viewed from the different chambers of the heart: perimembranous VSDs n=44

Table 31: Large PM VSDs n=38

Appearance of VSD- Large Perimembranous							
No	Age	Sex	Race	RV	LV 70	LV 40	Other
1.	0.67	M	Black	Mild infundibular stenosis.	-	-	-
2.	0.75	F	Black	-	-	-	Two small conal branches originating from RCA & cross RVOT.
3.	1.16	F	Black	Severe infundibular stenosis with PV thickening	Small ductal bump visible	Large PM VSD visible	R sided aortic arch. RCA gives rise to conal branch that crosses in area of RVOT.
4.	1.16	M	Black	Infundibular stenosis. R sided aortic arch.	-	-	L BT shunt noted filling PAs & is patent.
5.	1.17	F	Black	Severe infundibular stenosis	-	-	-
6.	1.33	F	Black	Tight infundibular stenosis with R to L shunting across VSD	-	-	Conal branch of RCA crossing region of RVOT
7.	1.5	F	Black	-	-	-	-
8.	1.5	M	White	Very narrowed infundibular segment. R to L shunt.	-	-	-
9.	1.5	F	White	Valvar stenosis with thickened leaflets.	-	-	-
10.	1.58	M	Colored	-	-	-	-
11.	1.58	M	Black	Large trabeculated RV. Tight infundibular stenosis. PV not thickened does not appear stenotic.	Large PM VSD. L sided aortic arch	-	No abnormal branch crossing RVOT.
12.	2	F	Black	Mild infundibular narrowing. RPA stenosed at origin.	-	-	Two small conal branches originating from RCA & cross RVOT.
13.	2	F	Colored	-	Large PM VSD with L to R shunting.	-	-
14.	2	M	Black	-	-	-	-
15.	2	M	Black	-	-	-	-
16.	2	M	Black	Tight infundibular narrowing. Thickened pulmonary valves	-	-	-
17.	2	M	Black	Large and trabeculated. RVOT appears to be divided into 2 separate narrow channels by longitudinal muscle band.	PM VSD with L to R shunting	-	-
18.	2	F	Black	Severe infundibular stenosis. PV thickened & domes in systole.	-	-	-
19.	3	F	Black	-	-	-	-
20.	3	M	Black	-	-	-	-
21.	3	M	Black	Tight infundibular stenosis.	-	-	-
22.	3	M	Black	RV well trabeculated. Narrowed segment separates inlet & apical portions of RV from distal infundibulum. No	Fairly large PM VSD with L to R shunt. Slight malalignment of	-	-

				infundibular narrowing. Typical appearance of double chambered RV.	aorta.		
23.	3	M	Black	PV appears thickened.	-	-	Azygos vein seen to join IVC below level of diaphragm. IVC appears to split in 2. One branch continuing as azygos vein to SVC/RA junction & other continuing as IVC to RA.
24.	4	M	Black	-	-	-	R sided aortic arch. LCA seen to arise from left posterior coronary sinus. RCA arises from right anterior coronary cusp. Conal branch crosses RVOT. Ductal bump on MPA
25.	4	M	Black	Infundibular stenosis/muscle band creating an infundibular chamber above stenosis.	-	-	-
26.	4	M	White	-	-	-	-
27.	4	F	Black	-	-	-	-
28.	5	F	Black	Infundibular area mildly stenotic.	-	-	-
29.	5	F	Black	-	-	-	-
30.	5	F	Colored	-	-	-	-
31.	5	M	Colored	-	Large PM VSD with L to R shunting across it. L sided aortic arch with minor degree of aortic override.	-	-
32.	5	M	Black	Large trabeculated RV with R to L shunting across VSD. Severe subclavian narrowing	-	-	Dominant LCA system with smaller RCA. Appears to be small conal branch arising from RCA crossing over RVOT.
33.	6	M	Black	Tight infundibular stenosis	Large PM VSD with aortic override	-	-
34.	6	M	Black	-	-	-	-
35.	6	M	Black	LV filling via VSD.	Aortic override with large PM VSD.	-	Intercostal vessels seen supplying the lung.
36.	9	F	Black	Very narrowed infundibulum. PV annulus hypoplastic. PV appears thickened & dysplastic. RV very hypertrophied.	-	-	RCA seems to arise from anterior R sided coronary sinus. Large conal branch crosses RVOT.
37.	9	F	Black	-	Large PM VSD with L to R shunting.	-	Absent LPA
38.	13	F	Black	Severe infundibular stenosis.	-	-	-

				Superior aneurismal dilation of LPA close to its origin.			

Table 32: Moderate PM VSDs n=3

Appearance of VSD- Moderate Perimembranous							
No	Age	Sex	Race	RV	LV 70	LV 40	Other
1.	2	M	Black	-	Moderately sized PM VSD.	-	Small conal branch crossing RVOT from RCA.
2.	2	F	Colored	Mild infundibular stenosis. R sided aortic arch	-	-	R coronary has conal branch that crosses RVOT.
3.	3	M	Black	Tight infundibular stenosis. Blind ended ductal bump noted on LPA	PM VSD	-	R sided aortic arch. Abnormal conal branch crossing RVOT from RCA.

Table 33: Small PM VSDs n=3

Appearance of VSD- Small Perimembranous							
No	Age	Sex	Race	RV	LV 70	LV 40	Other
1.	2	F	Black	Appears to be 2 separate channels from RV. Possibly subpulmonic VSD & RVOT. Double chamber RV with muscle band resulting in 2 channels	Small PM VSD visible. Aneurismal inter ventricular septum from attempted closure of VSD by TV. No coarctation of aorta.	-	-
2.	3	F	Black	-	Small PM VSD seen.	-	-
3.	5	M	Black	-	-	-	-

Table 34: Appearance of the VSDs when viewed from the different chambers of the heart: Outlet VSDs n=6

Appearance of VSD- Outlet							
No	Age	Sex	Race	RV	LV 70	LV 40	Other
1.	0.83	M	Black	-	-	-	-
2.	1.42	F	Indian	Narrowed RVOT in infundibular area. L to R shunting across VSD.	-	-	Small branch arising from circumflex that appears to cross region of RVOT anteriorly.
3.	1.5	F	Black	Mild infundibular stenosis	Subaortic VSD	-	-
4.	1.58	F	Black	Hypoplastic & short MPA. Infundibular short & narrow. PV thickened & domes in systole.	Large outlet VSD visible.	-	Large conal branch originating from RCA.
5.	6	M	Black	-	-	Small high subarterial VSD with L to R shunting. Aortic sinus on right appears squared off in keeping with prolapse of AV into VSD.	-
6.	7	M	Black	Tight infundibular stenosis	Large VSD noted	-	Anomalous R subclavian artery arising from descending aorta.

Great vessels of the heart

Table 35: Aorta and valve abnormalities: < 2 year age group n=15 (30%)

No	Age	Sex	Race	Ascend part (mm)	Arch			Valves (mm)				Descend part (mm)
					R/L	*% override*	*Size (mm)*	*AV*	*MV*	*TV*	*Details*	
1.	0.67	M	Black	15	Left	30	-	-	-	-	-	-
2.	0.75	F	Black	14.2	Left	50	-	-	-	-	-	-
3.	0.83	M	Black	-	Left	-	-	-	-	-	-	-
4.	1.16	F	Black	-	Right	20	-	-	-	-	-	-
5.	1.16	M	Black	-	Right	40	-	17	15	18	-	9.6
6.	1.17	F	Black	-	Right	50	-	-	-	-	-	pulsatile
7.	1.33	F	Black	14.7	Left	50	-	-	-	-	-	-
8.	1.42	F	Indian	Large	Left	35	-	-	-	-	-	-
9.	1.5	F	Black	20	Left	50	-	-	15	20	-	-
10.	1.5	M	White	-	Left	50	-	16	-	-	-	-
11.	1.5	F	Black	-	Left	30	-	-	-	-	-	-
12.	1.5	F	White	-	Right	-	-	-	-	-	-	-
13.	1.58	M	Colored	-	Right	-	-	TV leaflets thickened with abnormal looking fragile tissue of RV.				8
14.	1.58	M	Black	15	Left	20	-	-	-	-	-	-
15.	1.58	F	Black	-	Left	-	-	-	-	-	-	-

Table 36: Aorta and valve abnormalities: 2-4 year age group n=21 (42%)

No	Age	Sex	Race	Ascend part (mm)	Arch			Valves (mm)				Descend part (mm)
					R/L	*% override*	*Size (mm)*	*AV*	*MV*	*TV*	*Details*	
1.	2	M	Black	Large	Left	40	-	-	-	-	-	-
2.	2	F	Colored	-	Right	40	-	-	-	-	-	-
3.	2	F	Black	7	Left	50	-					
4.	2	F	Colored	Large	Left	50	-	16	18	12	-	-
5.	2	M	Black	15	Left	40	-	-	-	-	-	7.3
6.	2	F	Black	14	Left	-	-	-	-	-	-	-
7.	2	M	Black	Large	Right	50	-	20	-	-	-	-
8.	2	M	Black	Large	Left	50	-	-	13	15	-	8
9.	2	M	Black	17	Left	10	-	-	-	-	-	8.1
10.	2	F	Black	-	Right	40	-	-	14	12	-	-
11.	3	F	Black	-	Left	50	-	-	-	-	-	-
12.	3	F	Black	17	Left	10	-	-	-	-	-	-
13.	3	M	Black	15.3	Left	30	-	-	-	-	-	-
14.	3	M	Black	17	Right	30	-	12.6	8.9	-	-	14
15.	3	M	Black	16	Right	50	-	19	-	-	-	-
16.	3	M	Black	16	Right	35	-	16	19	15	-	-
17.	3	M	Black	12	Left	-	-	-	12	15	-	-
18.	4	M	Black	20	Right	87.5	-	-	-	-	-	9
19.	4	M	Black	17.6	Right	15.8	-	-	-	-	-	8.7
20.	4	M	White	-	Left	-	-	-	-	-	-	-
21.	4	F	Black	17	Left	50	-	20	12	18	-	-

Table 37: Aorta and valve abnormalities: > 4 years age group n=14 (28%)

No	Age	Sex	Race	Ascend part (mm)	Arch			Valves (mm)				Descend part (mm)
					R/L	*% override*	*Size (mm)*	*AV*	*MV*	*TV*	*Details*	
1.	5	F	Black	-	-	50	-	-	-	-	-	-
2.	5	F	Black	Large	Left	50	-	7	-	-	-	-
3.	5	F	Colored	Large	Right	50	-	-	-	-	-	-
4.	5	M	Black	Large	Left	50	-	-	-	-	-	-
5.	5	M	Colored	16	Left	10	-	-	-	-	-	-
6.	5	M	Black	-	-	-	-	-	-	-	-	-
7.	6	M	Black	-	-	-	-	-	-	-	-	-
8.	6	M	Black	Large	Left	50	-	-	-	-	-	-
9.	6	M	Black	15	Right	50	-	15	-	-	-	-
10.	6	M	Black	28	Left	55	-	29	14	28	Thickened TV septal leaflet + restrictive motion. Tri-leaflet AV.	10
11.	7	M	Black	25	Left	50	-	-	13	21	-	-
12.	9	F	Black	Large	Left	50	-	22	12	23	-	12
13.	9	F	Black	30	Right	50	-	9	-	-	-	10
14.	13	F	Black	-	Left	50	-	-	-	-	-	-

Table 38: Pulmonary vessels: < 2 year age group n=15 (30%)

No.	Age	Sex	Race	Pulmonary Trunk (mm)		Pulmonary Valve (mm)		Pulmonary Artery (mm)			
				Size	Des	Size	Description	MPA	RPA	LPA	Description
1.	0.67	M	Black	10.4	-	11	Mild infundibular stenosis	-	7	9	MPA had unusual appearance, proximal part dilated & then narrowed in supra valvar area.
2.	0.75	F	Black	5.4	-	9	Tight infundibular stenosis, some valvar stenosis. PV tricuspid with normal thin leaflets but mildly hypoplastic PV annulus. Small infundibular chamber present.	-	4.5	4.4	-
3.	0.83	M	Black	5	-	7	-	-	7	7	-
4.	1.16	F	Black	4.8	-	4	PV thickened & bicuspid, small PV annulus & infundibular stenosis. Infundibular os with fibrous edges near PV with hypertrophied infundibulum.	-	4.3	4.3	MPA short & hypoplastic.

5.	1.16	M	Black	-	-	10	Tricuspid	-	7	7.9	-
6.	1.17	F	Black	8	-	8	Small PV. Tight infundibular stenosis due to hypertrophied septal band & white stenotic like tissue in area of parietal extension of ventricular infundibular fold.	-	5	5	LPA stenosed
7.	1.33	F	Black	5.3	-	6.3	PV bicuspid, mildly hypoplastic annulus & supra valvar narrowing.	-	8	3	-
8.	1.42	F	Indian	9.1	-	6.5	Infundibular + supravalvar PS. PV bicuspid, mildly dysplastic with mild commissural fusion.	-	8.3	9.2	-
9.	1.5	F	Black	11	-	11	Severe infundibular stenosis. PV tricuspid, small infundibular valve underneath PV. Tight infundibular os surrounded by fibrous edges.	-	5.4	6.5	-
10.	1.5	M	White	5	-	9.4	Thickened PV leaflets dome in systole. Infundibular, valvar & supravalvar PS. PV bicuspid & dysplastic.	-	4.5	5.6	Narrowing of MPA at sino tubular junction.
11.	1.5	F	Black	-	-	-	PV tricuspid.	-	-	-	-
12.	1.5	F	White	4	-	4.5	Infundibular & valvar PS. Infundibular chamber long & narrow. PV annulus hypoplastic with dysplastic PV. PV bicuspid with thickened & dysplastic leaflets.	-	8	4	-
13.	1.58	M	Colored	12	-	12	PS: valvar & subvalvar level. PV leaflets dysplastic, pulmonary annulus hypoplastic. SS: level of infundibulum + hypertrophied muscle, no muscle bands to resect.	-	6.5	6.8	MPA hypoplastic
14.	1.58	M	Black	7.8	-	8.7	Infundibular PS. PV tricuspid with normal cusps.	-	5.2	6.7	-
15.	1.58	F	Black	-	-	-	Hypoplastic PV	-	5.3	-	Hypoplastic MPA

Table 39: Pulmonary vessels: 2-4 year age group n=21 (42%)

No.	Age	Sex	Race	Pulmonary Trunk (mm)		Pulmonary Valve (mm)		Pulmonary Artery (mm)			
				Size	Des	Size	Description	MPA	RPA	LPA	Description
1.	2	M	Black	8	-	6.5	Thickened PV + tight infundibular stenosis (os=3-4mm). Bicuspid.	-	6	5	MPA small & short.
2.	2	F	Colored	-	-	12	Tricuspid	-	10	10	-
3.	2	F	Black	4	-	5	PV dysplastic & practically atretic.	-	4	10	MPA hypoplastic.
4.	2	F	Colored	6	-	8	Supra valvar stenosis of 7mm. PV bicuspid & thickened.	-	6	7	-
5.	2	M	Black	16	-	-	-	-	8	8	-
6.	2	F	Black	12	-	-	Mild infundibular stenosis	-	10	8	-
7.	2	M	Black	11	-	-	Tricuspid, normal looking leaflets, mildly hypoplastic	-	5.5	5.5	-
8.	2	M	Black	5	-	8	Infundibular stenosis, supravalvular hypoplastic segment.	-	5.8	7	-
9.	2	M	Black	15.2	-	-	-	-	-	-	-
10.	2	F	Black	10	-	-	Severe infundibular PS. Thickened PV that domes. Bicuspid, nice leaflets, no commissural fusion	-	10	11	-
11.	3	F	Black	-	-	-	Bicuspid, thin leaflets, mild commissural fusion & mildly hypoplastic annulus.	-	-	-	-
12.	3	F	Black	-	-	-	-	-	-	-	-
13.	3	M	Black	-	-	9	Thickened, bicuspid & doming. PV annulus hypoplastic	-	10	15	MPA hypoplastic & short. LPA stenosed at origin, narrow over long segment.
14.	3	M	Black	7	-	9.5	Tricuspid	-	9	8	MPA small & underfilled
15.	3	M	Black	9.3	-	9	Tricuspid & thickened with commissural fusion	-	9	9	MPA small & short
16.	3	M	Black	16	-	16	-	-	8	9	-
17.	3	M	Black	-	-	-	-	-	-	-	-
18.	4	M	Black	9	-	8.3	Severe valvar PS + PR. Dysplastic	-	8	8	PA branches mildly hypoplastic, mild origin stenosis.
19.	4	M	Black	10	-	11	PV abnormally thickened	-	6	6	-
20.	4	M	White	-	-	-	Conus under PV, severe obstruction by fibromuscular tissue. 2 normal cusps, anteriorly no cusp only thick fibrous tissue	-	9	9	-
21.	4	F	Black	5.5	-	10	Infundibular + supravalvar PS, PV thickened. Z score for PV -1.	-	8	8	-

Table 40: Pulmonary vessels: > 4 years age group n=14 (28%)

No	Age	Sex	Race	Pulmonary Trunk (mm)		Pulmonary Valve (mm)		Pulmonary Artery (mm)			
				Size	Des	Size	Description	MPA	RPA	LPA	Description
1.	5	F	Black	-	-	-	Bicuspid, mildly thickened, several commissural fusion leaving a central orifice of 3mm.	-	-	-	-
2.	5	F	Black	12	-	10.5	Bicuspid, significant commissural fusion. Pulmonary orifice=5mm.	-	10	8	-
3.	5	F	Colored	5	-	4.5	Hypoplastic	-	10	Absent	MPA hypoplastic.
4.	5	M	Black	4	-	6.3	Dysplastic+ bicuspid	-	7	7	MPA hypoplastic
5.	5	M	Colored	7	-		Hypoplastic	-	11	11	MPA hypoplastic, supra valvar narrowing at level of sino-tubular junction.
6.	5	M	Black	5	-	-	-	-	10.1	5.7	-
7.	6	M	Black	-	-	-	-	-	-	-	-
8.	6	M	Black	13.7	-	-	-	-	4.7	5	-
9.	6	M	Black	6	-	7	-	-	5	6	-
10.	6	M	Black	5.6	-	12	Bicuspid & dysplastic, mildly fused commissures, shortened & thickened free edges. Sinotubular junction narrowed. Supravalvar PS=4.8mm	-	12	Absent	-
11.	7	M	Black	9	-	11	Bicuspid, normal looking leaflets.	-	10	10	MPA small.
12.	9	F	Black	14	-	8	-	-	7.8	9.8	-
13.	9	F	Black	10	-	16	Abnormal looking thickened leaflets, mildly fused commissures of PV + bicuspid	-	14	Absent	-
14.	13	F	Black	11	-	-	Eccentric & doming. Pulmonary bifurcation complex with strange poach. Bicuspid, thickened leaflets, eccentric commissural stenosis.	-	10	8.3	-

Cardiac catherisation data for Isolated VSDs

QP:QS	Systolic pulmonary artery pressures
2.3 : 1	55 mmhg
1.6 : 1	36 mmhg
2: 1	75 mmhg
2: 1	71 mmhg
2: 1	60 mmhg
4.4 : 1	45 mmhg
2: 1	53 mmhg
2.2 : 1	64 mmhg
-	-
1.3 : 1	87 mmhg
1.8 : 1	48 mmhg
1.5 : 1	37 mmhg
-	55 mmhg
-	59 mmhg
2: 1	55 mmhg
2.2 : 1	58 mmhg
2.5 : 1	30 mmhg
1.6 : 1	80 mmhg
2.3 : 1	39 mmhg
-	71 mmhg
2.6 : 1	65 mmhg
2.3 : 1	61 mmhg
3.1 : 1	42 mmhg
1.5 : 1	25 mmhg
0.9: 1	95 mmhg
1.3 : 1	83 mmhg
1.7 : 1	43 mmhg
3: 1	35 mmhg
4 : 1	35 mmhg
2: 1	40 mmhg
3.4 : 1	47 mmhg
0.9 : 1	20 mmhg
-	60 mmhg
1.9: 1	39 mmhg
4.6 : 1	98 mmhg
2.5 : 1	78 mmhg
2.2 : 1	40 mmhg
3.6 : 1	93 mmhg
2.7 : 1	95 mmhg
1.9: 1	45 mmhg
1.2 : 1	70 mmhg
3.3 : 1	75 mmhg
1.5 : 1	67 mmhg
0.6 : 1	83 mmhg
-	27 mmhg
1.9 : 1	85 mmhg
3.1 : 1	25 mmhg
-	-
2.5 : 1	80 mmhg
3.3: 1	50 mmhg

Cardiac catherisation data for TOF

QP:QS	Systolic pulmonary artery pressures
-	-
-	-
1: 1	-
-	-
-	-
-	-
-	-
-	-
0.3 : 1	-
0.5 : 1	-
2.4 : 1	50 mmhg
0.6 : 1	-
-	-
-	-
-	-
0.6 : 1	-
1: 1	38 mmhg
0.5 : 1	-
0.36: 1	-
-	-
0.86: 1	-
0.4 : 1	-
0.5 : 1	60 mmhg
-	-
0.2: 1	-
-	-
0.9 : 1	-
2.2 : 1	35 mmhg
-	-
0.3 : 1	-
0.6 : 1	-
0.5 : 1	-
-	-
0.6 : 1	-
-	-
-	-
-	-
1.7 : 1	28 mmhg
0.6 : 1	-
4.2 : 1	25 mmhg
0.23 : 1	-
-	-
DCRV	
QP:QS	Systolic pulmonary artery pressures
1.8 : 1	27 mmhg
1.8 : 1	30 mmhg
1.7 : 1	25 mmhg
1.9 : 1	26 mmhg
0.8 : 1	-
2 : 1	67 mmhg
1.2 : 1	41 mmhg
2.3 : 1	31 mmhg

Appendix B: Ethics clearance certificates

UNIVERSITY OF THE WITWATERSRAND, JOHANNESBURG
Division of the Deputy Registrar (Research)

HUMAN RESEARCH ETHICS COMMITTEE (MEDICAL)
R14/49 Ms M Osman

CLEARANCE CERTIFICATE	**M1204105**
PROJECT	An antomical and Retrospective Clinical Study of Interventricular Septal Defects (Previously M040921)
INVESTIGATORS	Ms M Osman.
DEPARTMENT	School of anatomical Sciences
DATE CONSIDERED	Ad hoc
+DECISION OF THE COMMITTEE*	Renewal Approved

Unless otherwise specified this ethical clearance is valid for 5 years and may be renewed upon application.

DATE 10/05/2012 **CHAIRPERSON** .....
(Professor PE Cleaton-Jones)

*Guidelines for written 'informed consent' attached where applicable
cc: Supervisor : Prof SL Cronje

DECLARATION OF INVESTIGATOR(S)
To be completed in duplicate and **ONE COPY** returned to the Secretary at Room 10004, 10th Floor, Senate House, University.
I/We fully understand the conditions under which I am/we are authorized to carry out the abovementioned research and I/we guarantee to ensure compliance with these conditions. Should any departure to be contemplated from the research procedure as approved I/we undertake to resubmit the protocol to the Committee. **I agree to a completion of a yearly progress report.**

PLEASE QUOTE THE PROTOCOL NUMBER IN ALL ENQUIRIES...

UNIVERSITY OF THE WITWATERSRAND, JOHANNESBURG

Division of the Deputy Registrar (Research)

HUMAN RESEARCH ETHICS COMMITTEE (MEDICAL)
R14/49 Osman

CLEARANCE CERTIFICATE	**PROTOCOL NUMBER M040921**
PROJECT	An Anatomical and a Retrospective Clinical Study of Interventricular Septal Defects
INVESTIGATORS	Miss M Osman
DEPARTMENT	School of Anatomical Sciences
DATE CONSIDERED	04.10.01
DECISION OF THE COMMITTEE*	Approved unconditionally

Unless otherwise specified this ethical clearance is valid for 5 years and may be renewed upon application.

DATE 04.11.11 **CHAIRPERSON** .....
(Professor PE Cleaton-Jones)

*Guidelines for written 'informed consent' attached where applicable

cc: Supervisor : Prof SL Cronje

DECLARATION OF INVESTIGATOR(S)

To be completed in duplicate and **ONE COPY** returned to the Secretary at Room 10005, 10th Floor, Senate House, University.
I/We fully understand the conditions under which I am/we are authorized to carry out the abovementioned research and I/we guarantee to ensure compliance with these conditions. Should any departure to be contemplated from the research procedure as approved I/we undertake to resubmit the protocol to the Committee. **I agree to a completion of a yearly progress report.**

PLEASE QUOTE THE PROTOCOL NUMBER IN ALL ENQUIRIES

University
of the Witwatersrand,
Johannesburg

Human Research Ethics Committee (Medical)
(formerly Committee for Research on Human Subjects (Medical)
Secretariat: Research Office, Room SH10005, 10th floor, Senate House • Telephone: +27 11 717-1234 • Fax: +27 11 339-5708
Private Bag 3, Wits 2050, South Africa

Ref: W-CJ-120411-1

11/04/2012

TO WHOM IT MAY CONCERN:

Waiver: This certifies that the following research does not require clearance from the Human Research Ethics Committee (Medical).

Investigator: **Department of Paediatrics (Head: Prof P Cooper).**

Project title: **Research on cadaveric material - Prof S Levin collection of cadaveric childrens' hearts.**

Reason: In terms of Chapter 8, sections 62-64 of the National Health Act No 61 of 2003 donated bodies and their tissues may be used for, among other purposes, health research. The Committee agrees that the hearts, donated by the parents of deceased children between 1965 and 1992 falls under the section above. The collection is housed in the Department of Paediatrics, Area 155 Charlotte Maxeke Johannesburg Academic Hospital. Research on the material is subject only to permission from the responsible person in the Department - the Head or person designated by the Head.

UNIVERSITY OF THE WITWATERSRAND
PROF PE CLEATON-JONES
HREC (MEDICAL)
2012 -04- 11
JOHANNESBURG

Professor Peter Cleaton-Jones
Chair: Human Research Ethics Committee (Medical)

copy: Anisa Keshav, Research Office, Senate House, Wits

Renew 2015

University
of the Witwatersrand,
Johannesburg

luman Research Ethics Committee (Medical)
ormerly Committee for Research on Human Subjects (Medical)
ecretariat: Research Office, Room SH10005, 10th floor, Senate House • Telephone: +27 11 717-1234 • Fax: +27 11 339-5708
ivate Bag 3, Wits 2050, South Africa

Ref: W-CJ-101109-1

09/11/2010

TO WHOM IT MAY CONCERN:

Waiver: This certifies that the following research does not require clearance from the Human Research Ethics Committee (Medical).

Investigator: **School of Anatomical Sciences (Head: Prof T J M Daly).**

Project title: **Research on cadaveric material.**

Reason: In terms of Chapter 8, sections 62-64 of the National Health Act No 61 of 2003 donated bodies and their tissues may be used for, among other purposes, health research. Use of such material is subject only to permission from the responsible person in the School of Anatomical Sciences - the Head or person designated by the Head.

UNIVERSITY OF THE WITWATERSRAND
PROF PE CLEATON-JONES
HREC (MEDICAL)
2010 -11- 09
JOHANNESBURG

Professor Peter Cleaton-Jones
Chair: Human Research Ethics Committee (Medical)

copy: Anisa Keshav, Research Office, Senate House, Wits

Subject:	Re: Fw: Ethics waiver to publish photographs of postmortem specimens
To:	mohseena@webmail.co.za
From:	Peter Cleaton-Jones <pcleatonjones@gmail.com>
Date:	Mon, 23 Jul 2012 15:50:34 +0200 (4 days, 23 hours ago)
Contacts:	pcleatonjones@gmail.com Add

Hi Mohseena,

I thought I'd replied to you. The waiver you have been given covers
the post-mortem photography.

Regards,

Peter Cleaton-Jones

On 23/07/2012, mohseena@webmail.co.za <mohseena@webmail.co.za> wrote:
>
> Sent from my BlackBerry® wireless device
>
> -----Original Message-----
> From: mohseena@webmail.co.za
> Date: Mon, 23 Jul 2012 10:30:31
> To: <pcleatonjones@gmail.co.za>
> Reply-To: mohseena@webmail.co.za
> Subject: Fw: Ethics waiver to publish photographs of postmortem specimens
>
>
> Sent from my BlackBerry® wireless device
>
> -----Original Message-----
> From: mohseena@webmail.co.za
> Date: Mon, 23 Jul 2012 10:20:57
> To: <pcleatonjones@gmail.co.za>
> Reply-To: mohseena@webmail.co.za
> Subject: Ethics waiver to publish photographs of postmortem specimens
>
> Dear prof
>
> My name is Dr. M. Osman. I contacted you 2 months ago. I am enquiring about
> whether or not I require a SPECIFIC WAIVER in order to PUBLISH PHOTOGRAPHS
> of postmortem specimens or will the waiver to UTILISE the postmortem
> specimen cover photography?
>
> My thesis underwent review and one of the examiners requests a re
> examination. One of the comments was the waiver for the photographs.
>
> I urgently request your assistance in this regard as my thesis is now ready
> for submission.
>
> Thank you prof
> M. Osman
> Sent from my BlackBerry® wireless device

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