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Children with Atrioventricular Septal Defects at Lagos State University Hospital

Adeola Animasahun, MD; Motunrayo Akinbami, MD; Olusegun Gbelee, MD; F. Bode-Thomas, MD; Adewale Oke, MD

Abstract

Background

Few studies in Nigeria and its region have described the pattern of congenital heart diseases. None of these studies have described the profiles of patients with Atrioventricular Septal Defect (AVSD). The aim of this study is to document the profile of patients with AVSD and the pattern of occurrence of cardiac and extracardiac lesions in this group of patients.

Methods

Prospective, involving consecutive patients with AVSD diagnosed at the Lagos State University Teaching Hospital (LASUTH), Nigeria between January 2007 and December 2014 as part of a large study. Data were analyzed using Microsoft Excel, supplemented by MegaStat software. Mean, standard deviation and other parameters were generated as necessary; p-values were derived. Statistical significance was set at p-value < 0.05.

Results

Atrioventricular septal defects were diagnosed in 7.4% of children with Congenital Heart Disease (CHD). The mean age at diagnosis was 12.99±6.11 months. Sixty-two-point-seven

percent (62.7%) of the subjects had no physical features suggestive of a syndrome, while 33.3% had clinical features of Down Syndrome. The mean age at diagnosis for non-syndromic children and children with Down Syndrome was 15.69±4.25 months and 5.28±0.75 months respectively (p=0.02). Isolated AVSD was found in 53.3% of the subjects; 9% had associated Tetralogy of Fallot (TOF).

Conclusion

Down Syndrome was the most common extracardiac anomaly associated with AVSD. In view of the development of pulmonary vascular disease early in children with Down Syndrome, we recommend routine screening of patients with features of Down Syndrome for the presence of Congenital Heart Disease to enable early intervention.

Keywords: Atrioventricular Septal Defect; Children; Lagos; Down Syndrome.

Introduction

Atrioventricular Septal Defect is a complex congenital heart defects that occurs from the abnormal development of the endocardial cushion. It accounts for 4 to 7.4% of all congenital heart defects.^{1,2}

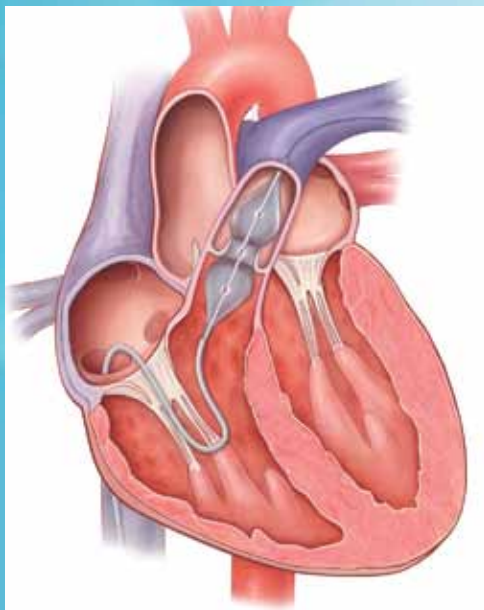
Atrioventricular Septal Defect occurs in 2 out of every 10,000 live births.³ It is the most common cardiac defect in children with Down Syndrome, accounting for 50% of cardiac defects in these children.⁴ About one third of all children born with AVSD have Down Syndrome.¹

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Atrioventricular Septal Defect is associated with other cardiac anomalies. In an interventional study by Pacifico et al⁵ on 29 subjects with complete AVSD, 41.3% and 34.4% had associated TOF and double outlet right ventricle respectively. In a retrospective study by Yildirim et al,⁶ double outlet right ventricle and right atrial isomerism were the most common cardiac anomalies associated with AVSD. TOF, dextrocardia and Transposition of the Great Arteries were also common anomalies seen.

Extra cardiac lesions can occur in up to 75% of cases with AVSD.² In a 10 year study by Huggon et al,⁷ Down Syndrome was the commonest karyotype abnormality, and was seen in 39% of the fetuses diagnosed with AVSD. Ivemark Syndrome, choroid plexus cyst, omphalocele, short femur and single umbilical artery are common extracardiac lesions identified in a retrospective study by Yildirim et al.⁶

The complete form of AVSD is more common in children with Down Syndrome, with the early and most severe form of progressive pulmonary vascular disease occurring if surgical intervention is not instituted in the first half of infancy.⁴ In a study involving 220 subjects with echocardiography diagnosis of AVSD by Marino et al,⁸ 47.7% of the subjects had Down Syndrome. The complete form of AVSD and TOF were more common in children with Down Syndrome than in other non-Down Syndrome subjects.

Few studies have described the pattern of Congenital Heart Diseases in Nigeria and the region.⁹⁻¹⁸ None of these studies have described the profiles of patients with AVSD; at best, the prevalence of AVSD has been mentioned,⁹⁻¹⁸ hence, the need for this study designed to document the profile of patients with AVSD and the pattern of occurrence of cardiac and extracardiac lesions in this group of patients.

Subjects and Methods

The study was prospective, involving consecutive patients with AVSD diagnosed using echocardiography at the Lagos State University Teaching Hospital (LASUTH), Nigeria between January 2007 and December 2014 as part of a large study. LASUTH is a tertiary institution that serves as a major referral centre in Nigeria.

Echocardiography was done by a Paediatric cardiologist according to the guideline for diagnosis.¹⁹ A GE Vivid E echocardiography machine with reference number 145021WP SN 2084 with appropriate-sized transducers was used.

All subjects had clinical evaluation done before echocardiography. Karyotyping was requested for children with features suggestive of chromosomal abnormalities. Clinical recognizable features were used for subjects that were unable to do karyotyping. Mongoloid slant, low set ears, epicanthic folds, flat facial profile and hypotonia which are chromosomal findings consistent with the diagnosis of Down Syndrome were features used to diagnosis Down Syndrome in subjects without karyotyping result.²⁶

Subjects' age, sex, age at diagnosis, indications for diagnosis and echocardiography diagnosis were inputted. Data was analyzed using Microsoft Excel program supplemented by MegaStat statistical package. Mean, standard deviation and other parameters were



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generated as necessary and p-values were derived. Statistical significance was set at p-value < 0.05.

Results

A total 315,150 children less than thirteen years old were seen as inpatients and outpatients in the paediatric department over the studied period. Out of these children, 1,495 had echocardiography done for various indications within the studied period. Among those who had echocardiography, seventy-five (6.57%) of the children with a structural heart defect had AVSD.

The mean age at diagnosis in subjects was 12.99 ± 6.11 months. The mean age at diagnosis was 11.22 ± 2.59 months in males and 11.46 ± 4.62 months in females as highlighted in Figure 1 ($p = 0.5$). The male to female ratio of subjects with AVCD was 3:2 ($p = 0.9$).

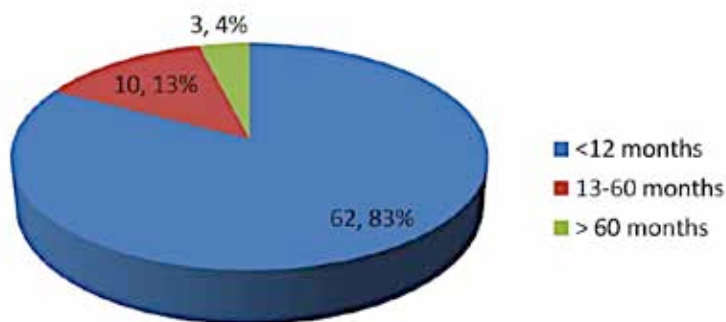


Figure 1. Age distribution of subjects.

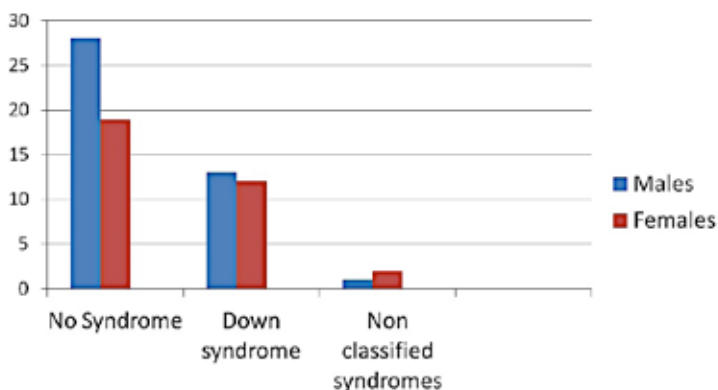


Figure 2. Frequency of syndromes in subjects according to their sex.

Table I highlights the indications for echocardiography among the subjects. The most common indications for diagnosis are suspicion of acyanotic congenital heart disease and Down Syndrome, which accounted for 42.7% and 26.7% respectively. A case of cerebrovascular accident with echocardiographic diagnosis of AVSD was seen in a 28-month-old boy who presented with central

Table I: Indications for Echocardiography in the Subjects

Indication	Frequency	Percentage
Breathlessness	32	42.7
Down Syndrome	20	26.7
Cyanosis	11	14.7
Failure to thrive	6	8.0
CCF	4	5.3
CVA	1	1.3
Dextrocardia	1	1.3
Total	75	100.0

cyanosis, fever, convulsions and right-sided hemiparesis. Many of the studied subjects (62.7%) had no physical features suggestive of a syndrome, while 33.3% of the studied subjects had clinical features of Down Syndrome. The mean age at diagnosis for non-syndromic children with AVSD and children with Down Syndrome with AVSD was 15.69 ± 4.25 months and 5.28 ± 0.75 months respectively ($p = 0.02$). There was a statistically significant difference in the age of diagnosis.

Table II shows the echocardiographic diagnosis of the studied subjects. AVSD with no associated cardiac anomalies occurred in 53.3% of the subjects, about 15% and 9% had associated secundum Atrial Septal Defect (ASD) and Tetralogy of Fallot respectively while 5.3% had dextrocardia.

Table II: Frequency of Other Cardiac Lesions Associated with AVSD in Subjects

Diagnosis	Frequency	Percentage
AVSD only	40	53.3
AVSD and ASD	11	14.7
AVSD and TOF	7	9.3
AVSD with PDA	6	8.0
AVSD, PS and Truncus arteriosus	5	6.7
AVSD with pericarditis	4	5.3
AVSD and TGA	2	2.7
Total	75	100

Discussion

In this study, 6.57% of children with CHD had AVSD. This finding is similar to the report of Mitchell et al,¹ Tantchou et al,¹² and Okoromah et al¹¹ where a prevalence of 7.4%, 7.3% and 7% were documented respectively, but lower than that of Sani et al¹⁸ among children from the northern part of Nigeria. The difference may be explained by the differences in the methodology and ages of the subjects in both studies. The Sani et al study included children and adults. It was over a shorter period of time (48 months), and involved fewer patients. Only 10 out of 122 patients with CHD had AVSD, hence, the finding in the current study, which was for a longer period (84 months) and involved more patients (75) with AVSD, is likely to be more



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representative. Moreover, the current study was carried out in the commercial capital of Nigeria (Lagos), which is a fair mixture of indigenes from various parts of the country, unlike the Sani et al study, which was carried out in the northern part of the country, and hence, is likely to be a reflection of the prevalence in the northern part of the country of AVSD, accounted for 0.02% of all children that presented to paediatric department of LASUTH, which is also similar to a report of 2 per 10,000 live births earlier documented by Colobro et al.³

The prevalence of AVSD in children with structural heart defect in this study is 6.57%. The lower prevalence in this study compares to a retrospective study by Sani et al¹⁸ in Kano, Nigeria with the prevalence of 8.2% may be explained by involvement of subjects up to 35 years in that study. The finding in this study is similar to a study by Tchoumi et al¹² in Cameroon with a prevalence of 7.3% despite the involvement of subjects up to 41 years old.

In this study, the most common indication for echocardiography was a suspicion of acyanotic congenital heart disease based on presenting symptoms of recurrent respiratory tract infection and murmur. Acyanotic cardiac defect being the commonest indication for echocardiography is in keeping with the findings of Geggel,²¹ in which evaluation of a murmur was the most frequent basis for cardiologist consultation; recurrent viral illnesses also accounted for a major indication for consultation.

The majority of the subjects (77.3%) were diagnosed after infancy with the mean age at diagnosis at 12.99±6.11 months. This is prognosticating because advance pulmonary vascular disease begins to develop in infancy with intima fibrosis occurring between 6-months to 1-year of age.²²

One of the findings in this study is the fact that about one-third of the subjects had Down Syndrome. A similar report was documented by Huggon et al.⁷ Many authors including Mitchell et al,¹ and Allas et al²³ documented AVSD as the most common CHD in children with Down Syndrome, although in some studies Ventricular Septal Defect was documented as being more common than AVSD in Down Syndrome.^{24,25} Down Syndrome is the most common syndrome identified in children with AVSD, accounting for 33.3% out of 42.6% syndromic AVSD in this study. The clinical presentations of mongoloid slant, low set ears, epicanthic folds, flat nasal bridge, and hypotonia were used in diagnosing Down Syndrome. These features are findings documented to be consistent in the diagnosis of Down Syndrome.²⁶

There can be an association of AVSD with other cardiac anomalies, such as PDA and TOF. In this study, 8% of the subjects had PDA; this is similar to an earlier report by Studer et al,²⁷ where PDA occurred in 10% of the AVSD cases. TOF is also an associated cardiac anomaly in AVSD; it was found in 9% of subjects in the current study. This is higher than 5% documented by Studer et al.²⁷ The reason for the higher proportion found in this study is not immediately clear. As it is, the case in this study, a high association of AVSD with TOF in Down Syndrome patients has been documented.²⁸ Seven subjects (9%) in the current study had AVSD associated with TOF of which six cases are children with Down Syndrome.

In conclusion, AVSD can occur in isolation or in association with a syndrome. Down Syndrome is the most common extracardiac anomaly associated with AVSD. The development of pulmonary vascular disease occurs in the first half of infancy with an earlier, and more severe form of the disease in children with Down Syndrome. There is the need for an early diagnosis and surgical interventions in infancy to prevent mortality that can occur from pulmonary vascular disease, hence, we recommend routine screening of patients with features of Down Syndrome for the presence of CHD.

Acknowledgement: We gratefully acknowledge the children who participated in this study and their parents.

Disclosures and Conflict of Interest: None

“Down Syndrome is the most common extracardiac anomaly associated with AVSD.... There is the need for an early diagnosis and surgical interventions in infancy to prevent mortality that can occur from pulmonary vascular disease, hence, we recommend routine screening of patients with features of Down Syndrome for the presence of CHD.”

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Pattern of Grown-Up Congenital Heart Disease in Lagos, Nigeria

By A.D. Olusegun-Joseph, MD; A.E. Okobi, MD; E.Y. Nkanor, MD; J.A. Mokwunyei, MD; F.E. Eto-Abasi, MD; A.C. Mbakwem, MD; J.N.A. Ajuluchukwu, MD

Introduction

The number of cases of Grown-up Congenital Heart Disease (GUCHD) is on the increase. This poses an added challenge to the cardiologist, and the health sector in general. The aim of this study was to assess the pattern of GUCHD in our centre; the types, frequency and complications.

Methods

This was a retrospective study of patients with Congenital Heart Disease (CHD), utilizing the echocardiography register from March 2001 to December 2010. The relevant data collected and analyzed included: age, sex, cardiac abnormalities and complications.

Results

A total of 6172 had echocardiography during the period under review. Of these, 143 patients (2.32%) had a diagnosis of GUCHD; between the ages of 12-80 years, with a mean age of 29.90 ± 14.74 years. The male to female ratio is 1.01:1 - 72 males (50.35%) and 71 females (49.65%). Most of the patients (97.9%) presented in their native, unoperated state, while only three patients (2.10%) were post-surgical. The most common cardiac anomalies were: Ventricular Septal Defect VSD, 55 (38.46%); Atrial Septal Defect (ASD) 51 (35.66%); and Tetralogy of Fallot (TOF) 14 (9.79%).

About 42% of patients had complications. Palpitation and breathlessness on exertion were the most common presentation.

Conclusion

The population with GUCHD in our study are predominantly young patients, with significantly high morbidity, presenting in their native, unoperated state. This population of people is on the increase worldwide, and we need to be prepared for particular challenges that they may have.

Keywords: Grown-up, congenital heart disease, morbidity, health challenges.

Introduction

Congenital Heart Defects (CHD) are abnormalities in cardiovascular circulatory structure and function that are present at birth.¹ They are the most common birth defects,² occurring in 0.5-0.8% of live births, 3-4% of still births; and 10-25% of abortuses.³ These heart

defects may be simple defects, or severe to complex, requiring immediate intervention at birth/ infancy and subsequent lifelong expert supervision and follow-up.^{1,3-5}

Most cases of CHD are diagnosed in infancy; however, despite recent advances in the diagnosis and treatment of congenital heart disease in infants, a significant number of patients are apparently missed during this phase and first present when much older.⁵⁻⁷ Grown-up Congenital Heart Disease patients, also known as Adult Congenital Heart Disease (ACHD), fall into the following categories: those who were diagnosed in childhood without surgical intervention with survival into adulthood, patients who were operated in childhood with survival into adulthood, and patients who are first diagnosed as adults.^{5,8} Although present at birth, many people with simple defects do not experience any symptoms until later childhood or adulthood.

The most common acyanotic congenital heart disease in the grown-up population is primarily left-to-right shunts, such as Ventricular Septal Defect (VSD), Atrial Septal Defect (ASD), Patent Ductus Arteriosus (PDA); while Tetralogy of Fallot is the most common example of cyanotic congenital heart disease.⁵ Other forms of CHD includes: Ebstein's Anomaly (EA), Transposition of the Great Arteries, dextrocardia, single ventricle hearts, uniaxial hearts, etc.

CHD patients require lifelong medical care.^{1,4-7} However, the challenge is that the grown-ups may be "lost" due to lack of transition from the pediatric cardiology clinic to the adult cardiologist in some cases. Furthermore, the special needs of GUCHD patients are also not well addressed by adult institutions, which are dominated by acquired heart disease, particularly hypertensive heart disease, dilated cardiomyopathy, valvular heart disease and coronary artery disease.⁵

The number of cases of GUCHD is on the increase globally, as adult survivors, reaping the gains of advancements in non-invasive diagnosis, early surgical and percutaneous intervention, now outnumber children in many developed countries.⁹ This poses an added challenge to the cardiologist and the health sector in general. Local studies on the subject are scanty, most coming from Europe and North America. This aim of this study was to assess the spectrum of GUCHD in our centre, and the peculiar clinical challenges that patients may present. The study will also help to sensitize the medical community to these important and largely correctable defects.

Methodology

This is a retrospective study of patients with CHD who had echocardiography performed

from March 2001 to December 2010; however, the records for year 2005 to 2007 could not be retrieved due to loss of data in the storage system. The data was obtained from our echocardiography register. Echocardiography studies were done using the Siemens Sonoline S1-450, and Aloka SSD 5000 in the Cardiovascular laboratory with a 3.5mHz transducer probe. Repeat scans were excluded. The procedures were done by consultant cardiologists, and Senior Registrars under supervision of consultants. Senior Registrars are resident doctors who undergo dedicated cardiology specialization training after passing the preliminary examinations (primaries and part I examination) of the medical postgraduate college.

Patients aged 12-years-old and above were recruited in the study. We recruited these patients because our hospital policy supports the transfer of patients aged 12- years-old-and-above to adult cardiology clinic, while still maintaining contact with their pediatricians. The aim is to facilitate smooth transition from the pediatric to the adult cardiology clinic.

The data collected included: age, sex, primary CHD diagnosis, secondary diagnosis, e.g. Hypertensive Heart Disease, and documented complications. Results were analyzed using the SPSS statistical package.

Approval for the study was obtained from the local Ethics Committee in the institution.

Results

A total of 143 patients with congenital heart defect were identified during the period of study. Their ages ranged from 12-80 years, with a mean age of 29.9 ± 14.74 years, of which 79% of the patients were < 40 years old; over 88% were < 50 years old (Table 1); 7.69% (11) of the patients were <16-years old. The oldest patient was an 80-year-old woman with bicuspid aortic valve and a diagnosis of heart failure. The oldest patient with VSD was a 76 year-old man, while the oldest patient with ASD was a 74-year old woman; both presented in heart failure. The oldest patient with TOF on the other hand was a

Table 1. Age Distribution		
Age group (years)	Frequency	Percentage (%)
< 20	34	23.78
20-29	47	33.57
30-39	32	22.38
40-49	13	9.09
50-59	5	3.49
≥ 60	12	7.69
Total	143	100

Table 2. Gender, Cardiac Defect Distribution				
	Sex		TOTAL	% of Total Population
	F	M		
VSD	28	27	55	38.46
ASD	26	25	51	35.66
TOF	7	7	14	9.79
PDA	4	3	7	4.89
Ebstein's Anomaly	0	4	4	2.80
DEXTROCARDIA	2	1	3	2.10
Endocardial Cushion Defect	1	2	3	2.10
Bicuspid Aorta	2	0	2	1.40
Cor Triatrium	1	1	2	1.40
Single Atrium	0	1	1	0.70
Sinus of Valsalva Aneurysm	0	1	1	0.70
Bicuspid Aorta	71	72	143	100

Legend: VSD - Ventricular Septal Defect; ASD- Atrial Septal Defect; TOF - Tetralogy of Fallot; PDA - Patent Ductus Arteriosus.

Table 3. Distribution of Age and Cardiac Defect in the Study Population							
Diagnosis	Age Group						TOTAL
	< 20	20-29	30-39	40-49	50-59	≥ 60	
ASD	12	23	5	3	4	4	51
Bicuspid aorta	0	0	0	0	0	2	2
Cor Triatrium (RV)	0	0	1	0	0	1	2
DEXTROCARDIA	0	0	1	1	0	1	3
Ebstein's Anomaly	0	1	3	0	0	0	4
Endocardial Cushion Defect	0	2	0	1	0	0	3
PDA	2	1	3	0	1	0	7
Single Atrium	1	0	0	0	0	0	1
Sinus of Valsalva Aneurysm	0	0	1	0	0	0	1
TOF	6	6	2	0	0	0	14
VSD	13	14	16	8	0	4	55
TOTAL	34	47	32	13	5	12	143

VSD- Ventricular Septal Defect; ASD - Atrial Septal Defect; TOF - Tetralogy of Fallot; PDA - Patent Ductus Arteriosus.

Table 4. Frequency Distribution of Complications		
	Frequency	Percent (%)
Pulmonary Hypertension	50	34.97
Dilated Chamber	24	16.78
Heart Failure	22	15.38
Eisenmenger	8	5.59
I.E	5	3.50
Thrombotic Risk	4	2.80
CVD	3	2.10

I.E = Infective endocarditis; CVD - Cerebrovascular accident. Please note that some patients had more than one complication.

34-year-old man with a documented history of Transient Ischemic Attack (TIA).

Seventy-two of the subjects were males (50.35%), while 71 (49.65%) were females. The gender/cardiac defect distribution is shown in Table 2. Most common cardiac anomalies were: VSD, 55 (38.46%); ASD, 51 (35.66%) and TOF, 14 (9.79%), with no significant gender differences in the anomalies. The age/ cardiac defect distribution is shown in Table 3.

Complications were noted in 60 (41.96%); with pulmonary hypertension 50 (34.97%), dilated chambers 24 (16.78%), and heart failure in 22

(15.38%), being the most frequent. Figure 1 and Table 4.

Discussion

There is a dearth of data on GUCHD in this part of the world. Even in western countries the precise data on the size and composition of the GUCHD population is still lacking, and can be assumed to change constantly.^{1,2,10} This study was borne out of the need to assess and highlight the spectrum of CHD in grown-ups in our centre; a major tertiary/ referral centre in South Western Nigeria, and compare with findings in other places.

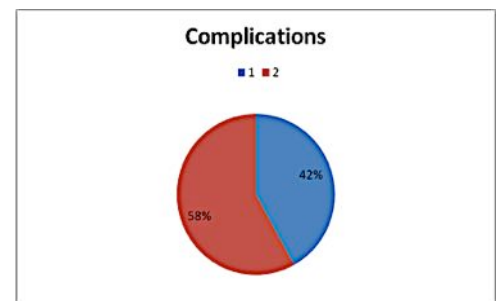


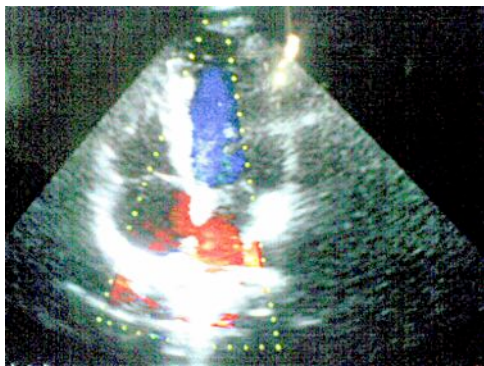
Figure 1. Presence of complications.

The burden of Congenital Heart Disease in Africa is quite significant, with up to 13% of new cardiac diagnoses in children and adults reported to be congenital.¹¹ In a resource challenged continent like ours the ripple effect is bound to be significant. Most of the patients in this part of the world present in their native form due to little or no availability/ affordability of cardiac surgical services in contrast to what entails in developed countries.¹¹⁻¹³ Furthermore, most of the patients with complex, severe defects hardly make it beyond early life stages.¹⁴ A similar trend was found in this study where the majority of the patients were natural survivors with diagnosis like VSD, ASD, TOF. Only three patients had had surgical correction, two patients with ASD, and one post TOF repair.

The relative frequency of CHD in this study is consistent with reports by other workers,^{15,16} with VSD, ASD and TOF being the most common defects in order of decreasing frequency. VSD and ASD are the most common acyanotic congenital heart defect in adults.^{5,6,17} The same trend is seen in pediatric studies.¹⁴ Okoroma et al in an earlier study done in our centre among the pediatric age group reported a similar trend, with VSD, ASD, TOF, ECD and PDA being the main lesions in decreasing frequency.¹⁴

The most common cyanotic heart disease is TOF, and this is consistent with findings by other workers.^{5,6,16} In our study only one patient of the lot had had surgical intervention; the others presented in their native, unoperated state. This is at variance with what entails in the developed countries where a sizable number of the patients would have had corrective surgery, thus offering them improved quality of life and survival opportunities.^{1,17}

Majority of patients in our study were in their second or third decade of life, with a declining prevalence in subsequent decades; there was however, a slight increase in the seventh decade compared with the sixth. About 79% of the study population were <40-years-old. Other investigators reported a similar trend in their study.^{15,16} This is quite disturbing considering the fact that these people are in their productive age group, forming a sizable portion of the work force of any society. The attendant morbidity and complications in this population will obviously reduce their quality of life and productivity. This may also be a reflection of the high mortality rate associated with these abnormalities in the



Picture 1. Apical four-chamber view showing Atrial Septal Defect (ASD) with blood flow from the left atrium to right atrium in a patient with Psoriasis.



Picture 2. Apical five-chamber view showing Ventricular Septal Defect (VSD) with blood flow from the left ventricle to the right ventricle.



Picture 3. Parasternal long axis view showing VSD with overriding aorta in a patient with Tetralogy of Fallot.

absence of early surgical correction.

There is age variation in the presentation of different defects, while the oldest patient with VSD was 76 -years-old, and ASD 74-years-old, the oldest patient with TOF was a 34-year-old woman. The age disparity shows the severity of the problem, and the attendant mortality. Morbidity and mortality are generally higher in cyanotic CHD compared with acyanotic CHD, ^{17,18} the case is even worse in a population like ours where most of the patients are natural survivors, yet to have surgical intervention. In contrast the oldest patient with TOF in the Euro Heart survey was 72-years-old.¹⁷ The age difference reveals the gains of early intervention, which a sizable number of that



Picture 4. Apical four-chamber view showing Ebstein's Anomaly. Note the apical displacement of the tricuspid valve and atrialization of the right ventricle.



Picture 5. Apical four-chamber showing severe tricuspid regurgitation in the patient with Ebstein's Anomaly.



Picture 6. Ventricular Septal Defect and overriding aorta in a patient with Tetralogy of Fallot.

study population benefited from. The need for proper development of manpower and cardiac surgical units cannot be overemphasized.

The development of complication is a major concern in GUCHD.^{1,19-20} In this study 42% of the patients had complications, with most of them having more than one complication. Pulmonary hypertension is a common complication in congenital heart disease and quite common in



Picture 7. Finger clubbing in a patient with Tetralogy of Fallot.

GUCHD.²¹ Its presence reduces the quality of life and survival of the patients.²⁰⁻²² Pulmonary hypertension was the most common complication in this study, affecting 50 (35%) of the study population. This is very high when compared with the UK national Audit, where the prevalence of Pulmonary Hypertension within the adult population was about 10%.²¹ The low prevalence in that study further underscores the gains of early diagnosis and intervention which majority of our patients, unfortunately, did not have.

The presence of secondary ailments in GUCHD has been documented by other workers.^{1,15} The GUCHD patient is not immune from other congenital and acquired diseases, and this can impact the morbidity and prognosis of the patients.^{23,24} The lesson here is that this population of patients should always have comprehensive evaluation, and there is usually need for multidisciplinary care.

In this study,²⁴ 16.8% of the population had documented secondary diagnosis. We acknowledge that there is likely to be a gross under-estimation of secondary diagnosis in the study considering the fact that we were not privy to full clinical history or case notes of the patients. Hypertension was documented in 6.99% of our patients, compared with the Euro Heart survey where 4% of the population had hypertension.¹⁷ In the study by Amaral et al,¹⁵ 9% of the unoperated arm had hypertension, as opposed to 18% in the operated arm. The comorbidities, including hypertension, in addition to requiring specific therapy, may hemodynamically affect the involvement of the underlying heart disease.

A major limitation of this study is that this is a hospital-based study, and may obviously not be a true reflection of the larger population of these patients. Furthermore, only those who had echocardiography were captured in the study. This is also a major limitation of the study.

In conclusion, the population with GUCHD in our study is predominantly young patients with significantly high morbidity, presenting in their native, unoperated state. The fact that most of the patients were yet to have any intervention contributed to the high prevalence of complications in this study, as progression of the pathophysiologic process is inevitable. We need to offer these patients early intervention to

reduce morbidity and improve their quality of life and survival.

Acknowledgement: We thank CNO Bastos for all her support and encouragement throughout this work.

Addendum

Secondary diagnosis were documented in 24 (16.8%) patients, commonest being hypertension 10(6.99%), Table 5. Others were Valvular Heart Disease (Mitral Regurgitation and Aortic Regurgitation; Mitral Stenosis and Mitral Regurgitation), Lutembacher Syndrome, Down Syndrome, etc.

Table 5. Secondary Diagnosis		
Secondary Diagnosis	Freq.	(%)
Hypertension	10	6.99
Mitral Valve Prolapsed	3	2.10
Valvular Heart Disease	2	1.40
Lutembacher Syndrome	1	0.70
COPD	1	0.70
Congenital Gynectresia	1	0.70
Hbss	1	0.70
Amyloidosis	1	0.70
Lupus Nephritis	1	0.70
Marfan's Syndrome	1	0.70
Down's Syndrome	1	0.70
Psoriasis	1	0.70
COPD - Chronic Obstructive Pulmonary Disease; Hbss - Hemoglobinopathy		

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Medical News, Products & Information

Compiled and Reviewed by Tony Carlson, Senior Editor

Edwards SAPIEN XT Valve Receives FDA Approval for Aortic Valve-In-Valve Procedures

PRNewswire -- Edwards Lifesciences Corporation announced it has received U.S. Food and Drug Administration (FDA) approval for aortic valve-in-valve procedures using the Edwards SAPIEN XT transcatheter heart valve.

"U.S. approval of the valve-in-valve procedure provides an important minimally invasive treatment option for patients who are at high risk for a subsequent open-heart surgery to replace their bioprosthetic valves," said Larry L. Wood, Edwards' Corporate Vice President, transcatheter heart valves.

At the October 27th *Transcatheter Cardiovascular Therapeutics (TCT)*, the annual scientific symposium of the Cardiovascular Research Foundation, in San Francisco, one-year patient outcomes were presented for patients treated with transcatheter SAPIEN XT valve implantation in surgical tissue valves. The valve-in-valve procedure was associated with a high overall one-year survival rate of 86.6%, and a low overall stroke rate of 3.7% in a very high-risk patient population, according to independently adjudicated data from the 197-patient PARTNER II Valve-in-Valve study. The study, which is a multicenter, non-randomized cohort of The PARTNER II Trial, included 97 patients enrolled in the primary registry, as well as 100 continued access patients.

"We were very pleased to see 100% survival at 30 days with the 100 high-risk patients treated with the SAPIEN XT valve-in-valve procedure in the continued access registry," said Danny Dvir, MD, interventional cardiologist at the Center for Heart Valve Innovation at St. Paul's Hospital, Vancouver, who presented the data. "This is quite remarkable and supports transcatheter aortic valve-in-valve replacement with the SAPIEN XT valve as a safe therapeutic alternative to reoperation for patients in need of a subsequent tissue valve replacement."

The Edwards SAPIEN XT valve was approved by the FDA in June 2014 for patients at high risk for native aortic valve replacement surgery, and received CE Mark for valve-in-valve procedures in early 2014. The SAPIEN valve platform has been used in the treatment of more than 100,000 patients worldwide. Additional company information can be found at www.edwards.com.

Cardiac Device Wearers Should Keep Distance from Smartphones

Cardiac device wearers should keep a safe distance from smartphones to avoid unwanted painful shocks or pauses in function, reveals research presented at *EHRA EUROPACE -- CARDIOSTIM 2015* by Dr. Carsten Lennerz, first author and

cardiology resident in the Clinic for Heart and Circulatory Diseases, German Heart Centre, Munich, Germany.

Lennerz said, "Pacemakers can mistakenly detect electromagnetic interference (EMI) from smartphones as a cardiac signal, causing them to briefly stop working. This leads to a pause in the cardiac rhythm of the pacing-dependent patient, and may result in syncope. For implantable cardioverter defibrillators (ICDs) the external signal mimics a life-threatening ventricular tachyarrhythmia, leading the ICD to deliver a painful shock."

Device manufacturers and regulatory institutions, including the U.S. Food and Drug Administration (FDA), recommend a safety distance of 15 to 20 cm between pacemakers or ICDs and mobile phones. The advice is based on studies performed primarily in pacemakers 10 years ago. Since then smartphones have been introduced and mobile network standards have changed from GSM to UMTS and LTE. New cardiac devices are now in use including: ICDs, cardiac resynchronisation therapy (CRT) and MRI compatible devices.

The current study evaluated whether the recommended safety distance was still relevant with the new smartphones, networks and cardiac devices. A total of 308 patients (147 pacemakers and 161 ICDs, including 65 CRTs) were exposed to the electromagnetic field of three common smartphones (Samsung Galaxy 3, Nokia Lumia, HTC One XL) which were placed on the skin directly above the cardiac device.

Lennerz said, "From earlier studies we know that the most vulnerable phases of a call are ringing and connecting to the network, not talking, so it was important to analyse these separately." More than 3,400 tests on EMI were performed. One out of 308 patients (0.3%) was affected by EMI caused by smartphones. This patient's MRI compatible ICD misdetected electromagnetic waves from the Nokia and HTC smartphones operating on GSM or UMTS as intracardiac signals.

Professor Christof Kolb, last author and head of the Department of Electrophysiology at the German Heart Centre, said, "Nearly everyone uses smartphones, and there is the possibility of interference with a cardiac device if you come too close. Patients with a cardiac device can use a smartphone, but they should not place it directly over the cardiac device. That means not storing it in a pocket above the cardiac device. They should also hold their smartphone to the ear opposite to the side of the device implant."

In a second study on EMI, researchers advise limiting exposure to high voltage power lines. The study was conducted in response to public concerns about bicycle routes and walking paths under high voltage power lines (230 kV and more), and whether these are safe for patients with cardiac devices. These high electric fields are also encountered in utility substations where employees who bring up power lines, conduct maintenance or work within the buildings (cleaners, for example) may be exposed.

Dr. Katia Dyrda, a cardiologist at Montreal Heart Institute, University of Montreal, said: "High electric fields may interfere with

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the normal functioning of cardiac devices, leading to the withholding of appropriate therapy (anti-bradycardia pacing, for example) or to the delivery of inappropriate shocks. The International Organization for Standardization says pacemakers and ICDs should give resistance up to 5.4 kV/m (for 60 Hz electric fields), but electric fields can reach 8.5 kV/m under high voltage power lines and 15 kV/m in utility substations."

She added, "There is a lot of interest in using the areas under power lines as bicycle paths or hiking trails because it's essentially free space. But patients and the medical community want to understand the risks. There are no recommendations from device manufacturers about power lines or higher electric fields." The study exposed 40 cardiac devices (21 pacemakers and 19 ICDs) from five manufacturers to electric fields up to 20 kV/m in a high voltage laboratory. The devices were mounted in a saline tank at human torso height. Devices were set up as both left- and right-sided pectoral implants.

The researchers found that when pacemakers were programmed to nominal parameters and in bipolar mode, they were immune to EMI up to 8.6 kV/m. But when programmed to higher sensitivity levels or in unipolar mode, the EMI threshold decreased to as low as 1.5 kV/m in some devices. When programmed to nominal parameters, all ICDs were immune to EMI up to 2.9 kV/m. There was no difference in EMI thresholds between left- and right-sided implants.

Dyrda said, "There is no significant concern for patients with pacemakers programmed in the usual configuration (nominal settings, in bipolar mode). For the minority of patients with devices in unipolar mode or with very sensitive settings, counselling should be given at implantation or at medical follow-up."

She added, "There is no need for patients with a pacemaker or ICD to avoid crossing under high voltage power lines (> 230 kV), but patients should avoid staying in a stationary position underneath them. Passing near pylons rather than between two pylons mitigates exposure to the electric field because the wires sag in the middle and the field is higher at this location."

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Visualize Radiation Exposure - Improve Patient Dose Management

By Allan Berthe, Contributing Editor, Special Projects

Radiation Conundrum

How much is too much? The long- and short-term effects of ionizing radiation to patients and clinicians are better understood today through study and research. The interventional clinician has the challenge to create the delicate balance of using enough radiation to produce a productive image, while limiting the exposure to the patient and to themselves. Over the last decade, interventional system providers have equipped systems with new technology to help manage radiation exposure. Hospital and clinician awareness supports an environment to conduct the safest possible procedure and strive to manage dose and follow the ALARA (As Low As Reasonably Achievable) principles. At a recent conference presentation, David Nykanen, MD, Arnold Palmer Children's Hospital, stated, "Of course the biggest problem is [that] you can't see radiation...the other issue is [that] tracking it is pretty hard." Now, new technologies are emerging to help manage radiation (while still producing a productive image), track amounts and provide ways to visualize the amounts of radiation exposure.

The Congenital Patient

Unlike older patients undergoing a single PCI procedure, a young congenital patient may be exposed to numerous procedures that utilize ionizing radiation to help diagnose and treat their condition. This radiation is cumulative and, therefore, should always be used prudently.

Because congenital conditions are discovered in a wide range of patients from newborns to mature adults, it is important to manage dose and take patient body-size into consideration (Figure 2).

Toshiba America Medical Systems' DTS (Dose Tracking System) technology tracks cumulative skin dose and displays it on a color-coded patient-specific graphic that is easily seen and read by the operator during interventional procedures.

Changing the Current Standard....What If?

Today interventional systems provide information related to amounts of dose being applied during the procedure. These are indicators as to how much dose is being used, but new technology offers



Figure 1. Picture of the Infinix-i interventional system with the DTS (Dose Tracking System) Display Screen on the larger format monitor.

improvements. Standard measurements displayed today include:

- Fluoroscopy Time....System timer that displays and sounds alarms (after every 5 minutes) the total amount of time that fluoroscopy is being applied during the procedure. Although this indicates how long fluoroscopy is being applied, it is not a precise indicator of how much is being applied, and what the skin dose is.
- Cumulative Air Kerma is a required system display and provides information on the total amount of radiation dose absorbed by air at a specific point in space relative to the X-ray source. This is measured free-in-air during an imaging or interventional procedure.
- Dose Area Product (DAP) provides measurement information for the entire amount of energy delivered by the X-ray beam, the measurement is derived from the area irradiated and the Air Kerma.

The amounts are displayed in numbers on the system screen that are interpreted by the clinician to provide an indication of radiation amounts. Although this information does aid clinicians, it does not tell the entire story regarding how much and where radiation is entering the patient's body. An important aspect of understanding the patient's exposure to radiation during the procedure is knowing amounts of skin dose. Clearly, a way to calculate and visualize the skin dose distribution and peak skin dose on a patient, is highly desirable.

The Advantages of DTS

Toshiba's proprietary technology allows clinicians to utilize standard forms of measurement / display and now add a real-time numerical display of peak skin dose



Figure 2. Sample of one of the patient size selection display screens.

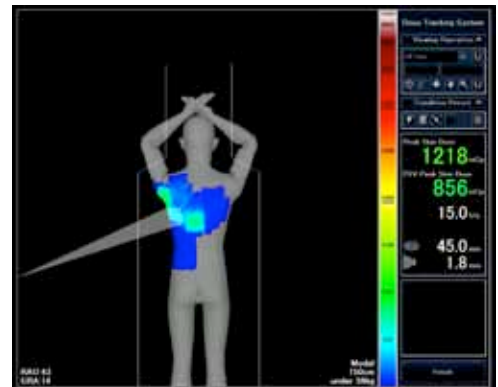


Figure 3. Example of the DTS operation screen display, shows direction of X-ray beam and where radiation has been applied, along with graphical and numerical indication of dose estimates.

and FOV (field-of-view) peak skin dose. These are displayed on a graph and an anatomical model to show skin dose distribution. This advantage aids clinicians to make real-time decisions regarding dose application and includes:

- The preselecting of patient-size from anatomical model menu to match actual patient size enhances source to skin distance corrections.
- The ability to track and include movement of the X-ray beam in relationship to the anatomical model to provide a distribution of dose rather than a single cumulative number.

- The taking into account of other procedure room influences such as patient table support, tissue absorption and backscatter characteristics.

This comprehensive graphical scale and patient anatomical model displays how much dose is being applied to general and specific areas, and allows clinicians to make real-time changes to system parameters and projection angles to alter radiation exposure amounts. This amounts to a visual early warning system that enhances patient and staff safety and provides clinicians the opportunity to effectively manage deterministic radiation risk.

Real DTS Application

Example of the DTS operation screen display, shows direction of X-ray beam and where radiation has been applied, along with graphical and numerical indication of dose estimates (Figure 3).

DTS has made its way into the market place, and is currently being used at leading congenital hospital Nationwide Children's Hospital, in Columbus, Ohio. John Cheatham, MD, FSCAI, has been utilizing the technology for several months and realizing benefits. To enhance the visualization performance, the system has a patient size-selection menu to facilitate utilization on a wide range of patient sizes.

Dr. Cheatham shared, "We provided Toshiba input on the patient selection menu, and our team values the ability to select different patient sizes to accommodate our wide range (from newborns to adults) of patients." This technology, allowing the visualization of radiation application to the patient, is another step the industry is taking to enhance safety and overall dose management. Regarding this, Dr. Cheatham indicates, "I appreciate the system's ability to track the X-ray beam and C-arm movement while providing an immediate real-time, color-coded visual que related to patient radiation exposure."

CCT

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CHIP NETWORK

CONGENITAL HEART PROFESSIONALS

WHAT IS THE CHIP NETWORK? - The CHIP Network, the Congenital Heart Professionals Network, is designed to provide a single global list of all CHD-interested professionals in order to:

- Connect pediatric and adult CHD-interested professionals to events, conferences, research opportunities and employment
- Keep members up with the literature through the monthly *Journal Watch* service
- Increase education and provider awareness of new developments
- Bring the pediatric and adult congenital heart communities into closer contact
- Offer a communication tool for critical issues

WHO SHOULD PARTICIPATE? - The CHIP Network is all inclusive and is comprised of everyone who considers themselves a congenital heart professional or administrator, including: Pediatric cardiologists, ACHD cardiologists, RNs and APNs, Cardiac surgeons, Cardiac care associates, Trainees/fellows, Administrators, Psychologists and Mental health professionals, Researchers/scientists, Intensivists, Anesthetists, Industry representatives

OUR SUPPORTING PARTNERS:

- Adult Congenital Heart Association
- Asia Pacific Society for ACHD
- Children's Hospital of Philadelphia Cardiology meeting
- Cincinnati Children's Hospital
- Congenital Cardiology Today (official publication of the CHIP Network)
- Congenital Heart Surgeons Society
- ISACHD
- Japanese Society of ACHD
- Johns Hopkins All Children's Heart Institute
- North American ACHD program
- Paediatric Cardiac Society of South Africa
- Pan Arab Congenital Heart Disease Association
- PCICS
- PICS
- Specialty Review in Pediatric Cardiology
- World Congress of Pediatric Cardiology and Cardiac Surgery

JOIN US - Membership is Free!

The CHIP Network management committee invites the participation of other organizations who want to communicate with all or some of the congenital heart professionals on this list. Please contact Dr. Gary Webb (gary.webb@cchmc.org) to ask that your organization's or institution's name be added to the list of partner organizations.

Register at: www.chipnetwork.org.



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