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CARDIAC CATHETERIZATION IN GROWN-UP PATIENTS WITH CONGENITAL HEART DISEASES – INDICATIONS AND COMPLICATIONS

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Contribution

All the authors contributed significantly to the research that resulted in the submitted manuscript.

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ABSTRACT

Background: Congenital heart defects are the most common congenital anomalies with incidence of 1% live births. The number of grown-up patients with congenital heart defects is growing worldwide including Pakistan, with proportionate increase in the number of diagnostic as well as interventional cardiac catheterization procedures.

Objective: The aim of this study was to evaluate the indications and immediate complications of cardiac catheterization in GUCH.

Methodology: This descriptive study was conducted in the Department of Pediatrics from December, 2010 to June, 2012. All adult patients (age more than 18 years) were included in the study, who underwent cardiac catheterization for congenital heart defects. Data analysis was computer-based using SPSS 17 version and descriptive analyses were applied where appropriate.

Results: A total of 208 cases were included with mean age of 28.7 ± 10.3 years and 56.7% were female. 105 cases were therapeutic catheterizations and 103 cases underwent catheterization for diagnostic work-up. Therapeutic procedures included atrial septal defect device closures (ASD) (n=65) with 92.3% success rate, and patent ductus arteriosus device closure in 18 patients with 100% success. Of the diagnostic catheterizations, tetralogy of Fallot, as a group (n=30), was the most common indication. Complications encountered were transient arrhythmias in 4 patients, failure to deploy ASD device in 5 cases and local groin hematoma in one case.

Conclusion: The most common indications of cardiac catheterization in GUCH are ASD device closure and diagnostic cardiac catheterization for TOF with complication rate of less than 5%.

Key words: Congenital heart defects; Atrial septal defect; Tetralogy of Fallot, Patent ductus arteriosus.

INTRODUCTION

Congenital heart defects (CHDs) are the most common congenital anomalies, reported around 0.8-1% of live births.¹⁻³ Due to non-structured health system in Pakistan, large numbers of children with CHDs are deprived of definitive cardiac surgeries, resulting in increasing number of adults with uncorrected heart defects. This group is commonly known as grown up patient with CHD (GUCH) and carries numerous inherent complications including cardiac failure, infective endocarditis, arrhythmias, pulmonary hypertension(HTN) and cerebral events.⁴ Maternal mortality is alarmingly high in GUCH women.⁵ An important aspect of GUCH is regarding delineation of surgical anatomy and decision regarding operability in the setting with pulmonary HTN, thus necessitating cardiac catheterization to reach a viable decision. Another important issue is interventional catheterization in GUCH, which is increasingly becoming common, due to increasing number of available devices/balloons and stents for percutaneous transcatheter intervention as well as increasing awareness in patients along with fear of major cardiac surgeries. To the best of our knowledge, there is no study has yet been conducted in Pakistan, addressing the issues of cardiac catheterization in GUCH especially its indications and associated complications. This prospective study was specifically undertaken with the aim to determine the indications and complications of catheterization in GUCH at our institution.

METHODOLOGY

This descriptive study was conducted in the department of Pediatric Cardiology at from December 2010 to June 2012 after approval of Institutional Review Board and Ethical committee. A total of 208 consecutive adult patients (age more than 18 years) were included in the study, who underwent cardiac catheterization (either diagnostic or therapeutic) for CHD during the study period. Patients were selected to undergo either therapeutic or diagnostic catheterization, on the basis of clinical presentation and echocardiographic evaluation. A short history and focused clinical examination was performed for each case followed by echocardiography a day before cardiac catheterization study. Data was collected on specifically designed proforma and all information of

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the catheterization study was collected for each of the 208 patients. Demographic variables such as age, gender, height and weight were recorded for each case. All patients were admitted on the day of cardiac catheterization and were kept nil per mouth for at least six hours prior to the start of procedure. After sedation with local anesthesia or general anesthesia in some cases, vascular access was established and strict aseptic techniques were followed. Procedures were either percutaneous transcatheter interventions or diagnostics catheterization studies. Diagnostic catheterization were divided into cyanotic and acyanotic groups. At the end of catheterization, type of procedure, results, total procedural time, fluoroscopy time, amount of contrast used, details of therapeutic interventional devices or balloons/stents, oxygen saturations and pressure recordings, mode of anesthesia, vascular access details (arterial, venous or both) and any complications during or after the procedure were recorded for each case. All cases were kept under observation for next 18-24 hours in post cath wards.

All the data was entered in SPSS version 17 (predictive analytics software and solutions) and data analysis was computer-based and descriptive analyses were applied as appropriate. Since the study was descriptive and involved no comparisons, therefore, no test of significance was applied.

RESULTS

A total of 208 cases were included in this study with mean age of 28.7 ± 10.3 years (Range 19 - 73 years). Males patients were 90 (43.3%) while 118 (56.7%) were females. The mean weight of patients was 53.6 \pm 11.1 kg (32-94 kg) and mean height was 160.4 ± 8.5 cm (134 - 182 cm). Mean procedural time was $36.9 \pm$ 16.8 minutes (12 – 150 min) while mean fluoroscopy time was 10.7 ± 9.7 min (1.5–91 min). Of the total procedures done, 181(87%) were done under local anesthesia while in 27(13%) patients general anesthesia was used. Regarding access point, in 113(54.3%) patients both arterial and venous access were used for cath study. In 81 (38.9%) patients only venous access was used while 14 (6.7%) patients had only arterial access used for catheterization. 105 cases were therapeutic catheterizations and 103 cases underwent catheterization for diagnostic work up only. Table 1 shows the indications of cardiac catheterization in study population.

Percutaneous transcatheter intervention group included 105 adult patients (69 being females) underwent percutaneous transcatheter intervention in our study with mean age of 31.2 years. The largest numbers of therapeutic procedures were atrial septal defect (ASD) device closure, accounting for 65 (61.9%) cases with 92.3% success rate (Fig 1a). In five cases, ASD device closure was unsuccessful as multiple attempts failed to deploy the device in satisfactory position.

Other therapeutic procedures included patent ductus arteriosus (PDA) device closure in 18 (17.1%) patients with 100% success (Figure 1b), pulmonary valve ballooning in 9 cases and ballooning /stenting for Coarctation of aorta in 5 patients (Table 1, Figure 2). The longer procedural and fluoroscopic time was in major aorto-pulmonary collateral artery (MAPCA) coiling cases, mean of 73 and 34 minutes respectively. During PDA device closure, snare was used as wire could not cross the duct from venous side, in 4 cases.

Of the diagnostic catheterizations, tetralogy of Fallot (TOF) as a group (including classic TOF, TOF with

absent pulmonary valve, TOF palliated with Aortopulmonary shunts) was the most common indication which accounted for 30 (29.1% in diagnostic group) cases (Figure 3 a&b).

ASDs with pulmonary hypertension (also including partial ASDs and two cases with ASD patch leak) and ventricular septal defect (VSD) with pulmonary hypertension were the next most common indications for diagnostic catheterizations, in 27 (26.2%) and 10 (15.5 %) patients respectively. 16 cases with mid right ventricle (RV) band with VSDs or ASDs were also included in diagnostic group (Table 1). The complex catheterizations included unbalanced Atrioventricular, transposition of great arteries (TGA), congenitally corrected TGA (CcTGA), Truncus arteriosus, total anomalous pulmonary venous return (TAPVR) and single ventricular physiology as double inlet left ventricle. The mean age in diagnostic group was 26.2 years. The diagnostic catheterization in age group 19-30 years was 55.8% in comparison to 38.3% in age group 31-45 years and only 21.4% in cases above 46 year of age.

Diagnostic catheterization	Numbers (% in group)	Interventional procedures	Numbers (%in group)
TOF/Pulmonary atresia	30 (29.1)	ASD device closure	65 (61.9)
ASD + pulmonary HTN	27(26.2)	PDA device closure	18 (17.1)
Mid RV band	16(15.5)	Pulmonary valvuloplasty	09 (8.6)
VSD + Pulmonary HTN	10 (9.7)	Coarctation stenting	05 (4.8)
PDA + Pulmonary HTN	03 (2.9)	Aortic valvuloplasty	02 (1.9)
Unbalanced AVSD	02 (1.9)	MAPCAs coiling	05 (4.8)
Others /complex	15 (14.6)	Device BT shunt	01 (1)

Table 1: Numbers and percentage of cases according to diagnostic or interventional procedures

Figure 1a & b: ASD & PDA device closure in adult patients





Figure 2: Stenting of adult female patient with re-coarctation

Figure 3 a & b: RV out flow is opacified & MAPCAs are seen arising from upper thoracic aorta in TOF adult patient



Complications encountered in the study were transient arrhythmias in four patients, failure to deploy ASD device in five cases and local groin hematoma in one case. No mortality, cardiac perforation or local vascular access site complication occurred.

DISCUSSION

In our setup, pediatric cardiologists are the ones, dealing with both diagnostic as well as therapeutic cardiac catheterization in GUCH, having vast experience of adults as well as pediatric catheter studies and percutaneous transcatheter interventions. In Pakistan, the number of children born each year with CHDs are roughly 20 times than total number of cardiac surgeries performed all over the country. This gap is surgery and if they somehow survive to adulthood, the result is GUCH. With increasing number of GUCH cases, the numbers of catheterization studies have also increased in parallel, both diagnostic as well as therapeutic, nearly 50% of each group in our study. In our study, most patients were female which is against the reported trends in studies of pediatric age groups.^{1,6} This highlights another sore point, as parents are more concerned with the health of their young male off springs in a male dominant society like ours. Females are brought to health facilities around marital age or even after the marriage, when cardiac status complicates the pregnancy. Nevertheless, some CHDs present in adulthood like ASD, which was about 31% of

increasing each year leaving many children with no

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(n=66) total cases in our study, with mean age of 34 years and in 5 cases ASD device cloud not be deployed (in spite of detailed pre and per procedural transesophageal echocardiography). These were referred for surgery. Main reasons of failure were floppy rims in 3 cases, inadequate IVC rim in 1 case and an additional small ASD distant to the large ASD in 1 case. Multiple attempts were needed in 10% cases before successful ASD device closure but no device dislodged after deployment (Fig 1a). The near similar results are recently reported by Vijarnsorn et al in their study of children and adults undergoing ASD device closure, with about 93% success rate, 4% failure and 1% device embolization.⁷ Another group was of PDA device closure, accounting for 8.6% of total patient population with 100% success (Fig 1b). Adult PDA carries risk of about 2% mortality per year⁸ and PDA device closure safety is widely reported in literature.^{8,9}

In four cases, it was difficult to cross the wire from PA to Aorta through PDA especially in small PDAs; snare was used in these cases. Before proceeding for PDA device closure, cases were subjected to catheterization and after determining pulmonary vascular resistance, intervention was undertaken. In three cases, PDA was occluded with the appropriate size balloon, to determine response of pulmonary artery pressures before suitability for device. No case was declared inoperable on the basis of catheterization data. In our observation, the adults PDA device closure is more time consuming than pediatric patients, along with increased fluoroscopic time. Other interventions included balloon/stenting group such as pulmonary valve stenosis (n=9), aortic valve stenosis (n=2) and Coarctation of aorta stenting (n=5) with 100% success (Fig 2). Five patients underwent coiling of MAPCAs, followed by total correction. The efficacy of balloon pulmonary valvuloplasty is beyond any doubt and at the same time it is safe with careful technique. We achieved at least 50% reduction in peak to peak pressure gradient across pulmonary valve in all cases with 2 cases of transient rhythm disturbance during ballooning. In a study from Taiwan, authors reported safety in an 80 year old patient as well.¹⁰

In diagnostic group, cyanotic CHDs were about 46% and acyanotic CHDs 54%. Commonest indication for diagnostic cardiac catheterization was TOF including a case of TOF with absent pulmonary valve syndrome (APVS), eleven BT shunts, one pulmonary atresia and 17 cases with TOF with pulmonary stenosis. Catheterization study was considered complete with contrast angiography of left ventricle, RV, RV outflow, branch pulmonary arteries, Aorta along with clear delineation of origin & course of coronary arteries/systemic veins and presence or absence of MAPCAs (Fig 3 a & b). It needed around one hour time to complete TOF study in most cases along with high volume of contrast, increased fluoroscopic time and thus higher amount of radiation to patient as well as staff in comparison to pediatric population. In these patients, CT angiography is an alternative but with disadvantage of no hemodynamic data. MRI is promising, not available with us yet. Delaying corrective surgery, not only subject patients to hazards of cardiac catheterization but with increased mortalities and residual defects in adults with TOF. In our study, eleven adult TOF patients were palliated with aorto-pulmonary shunts and cardiac cath in these cases was suggestive of multiple collaterals in all and functional shunts in six cases. The quality of life is certainly compromised where total correction is delayed to adulthood. It is worth mentioning here that diagnostic cardiac catheterization in simple TOF is a rarity in developed countries, primarily due to two reasons. Firstly the age of total correction is in early infancy in developed countries with advantage of better echocardiographic assessment at young age along with better noninvasive diagnostic modalities including MRI. Unfortunately, both of these factors are lacking in our country.

In 27 cases (60% females), cardiac catheterization was performed with provisional diagnosis of ASD and pulmonary HTN. In all cases shunt across ASD was left to right, but with mean pulmonary artery pressure of more than 25 mmHg. The presence of pulmonary HTN associated with ASDs is reported and especially sinus venosus type ASD.¹¹ Idiopathic or primary pulmonary hypertension was diagnosed with ASD kept open and treatment with calcium channel blockers were given, if evidence of vasoreactivity was present. Other cases with left to right shunts included VSDs & PDA, where catheterization study was simple and near 90% cases were declared operable, but it took higher amount of time and radiations in comparison to young pediatric patients. Sixteen cases with mid RV band (14 with VSD & 02 with ASD) were also subjected to catheterization. study proved fairly simple with no complications. Though it is not necessary to do cardiac catheterization in all cases of mid RV band, but there are number of cases where angiography may be mandatory to clearly delineate the anatomy of RV and LV and to rule out defects like VSD before proceeding for surgery. Local vascular complications in our study were groin hematoma in one case, which was treated conservatively. Local vascular complications reported by Chue *et al* in their retrospective study of 197 procedures in adult CHDs were 3.6%.¹² Frequency of systemic complications (4.3%) and local vascular complications (<0.5%) were also considerably low as compared to 11% as reported by Mori *et al*¹³ and comparable to that reported by Tavli *et al* in their study of pediatric population.¹⁴

CONCLUSION

The most common indications of cardiac catheterization in grown up patients with CHDs are ASD device closure followed by diagnostic cardiac catheterization for TOF. In experienced hands, it is safe to perform catheterization studies in these cases, however, with prolonged procedural and fluoroscopic times.

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