AN EXPERIENCE OF CARDIAC CATHETERIZATION PROCEDURES IN PATIENTS WITH INTERRUPTED INFERIOR VENA CAVA

Mehboob Sultan, Khurram Akhtar*, Vaqar Elahi Paracha, Nadeem Sadiq*, Imran Bashir, Amjad Mehmood*, Khush Bakht Khan Awan

Army Cardiac Centre, Lahore Pakistan, *Army Forces Institute of Cardiology/National Institute of Heart Disease (AFIC/NIHD)/National University of Medical Sciences (NUMS) Rawalpindi Pakistan

ABSTRACT

Objective: Interrupted inferior vena cava (IVC) is a well-recognized but a rare congenital anomaly due to absence of hepatic portion of the IVC and can therefore pose problems while undertaking cardiac catheterization in these particular patients. We are sharing our experience of cardiac catheterization in patients with interrupted IVC.
Study Design: Retrospective descriptive study.
Place and Duration of Study: Armed Forces Institute of Cardiology/National institute of heart diseases (AFIC/NIHD) Rawalpindi, from Jan 2017 to Dec 2018.
Methodology: This retrospective descriptive study was done at the Paediatric cardiology department of the Armed Forces Institute of Cardiology/National institute of heart diseases (AFIC/NIHD) Rawalpindi and comprised of 34 consecutive cases with interrupted IVC, who underwent cardiac catheterization procedure from January 2017 to December 2018.
Results: Total of 34 consecutive patients with interrupted IVC underwent cardiac catheterization procedure (29 diagnostic procedures and 5 interventional procedures) with mean age of 4.3 ± 4.5 years, including 19 males & 15 females. The mean procedural time and fluoroscopy time were 55 & 17 minutes respectively and no major complications were encountered in study population.
Conclusion: Cardiac catheterizations in patients with interrupted IVC can be challenging especially in device closure of atrial septal defects and patent ductusarteriosus.

Keywords: Cardiac catheterization, Interrupted inferior vena cava.

This is an Open Access article distributed under the terms of the Creative Commons Attribution License (http://creativecommons.org/licenses/by/4.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

INTRODUCTION

Interrupted inferior vena cava (IVC) is a rare but well-recognized congenital vascular anomaly that results due to absence of hepatic portion of the IVC. It commonly leads to drainage of blood from the caudal half of the body alternatively through the azygous veins, hemiazygous or accessory hemiazygous veins to superior vena cava (SVC). Isolated anomaly is generally asymptomatic and may occur in about 0.3% of the population[1]. However, when in association with congenital heart diseases predominantly cardiosplenic syndrome (heterotaxy) and left isomerism, its incidence is high[1-4]. Albeit, easily diagnosed on transthoracic echocardiogram, it is typically an incidental finding when failure to obtain vascular access to right heart occurs from femoral vein. It is also an uncommon cause of DVT (deep venous thrombosis)[1,3] especially where no cause is established.

During embryogenesis, the cardinal veins namely the common cardinal veins, anterior cardinal veins, posterior cardinal veins, supracardinal and the subcardinal veins constitute the main venous drainage system of the embryo. The ultimate venous system is formed by a series of modifications of these veins. The IVC also forms during these changes and is composed of four main segments[5].

- A hepatic segment derived from hepatic veins (proximal part of right vitelline vein) and hepatic sinusoids.
- A pre-renal segment derived from the right subcardinal vein.
- A renal segment derived from the subcardinal-supracardinal anastomosis.

Correspondence: Dr Mehboob Sultan, Department of Paeds Cardiology, Army Cardiac Centre, Lahore Pakistan
Email: drmehboobsultan@gmail.com

- A post-renal segment derived from the right supracardinal vein.

Since the process of formation of SVC and IVC is complex, variations in the mature form may occur such as double superior and inferior vena cavae, left sided inferior vena cava and interruption of abdominal course of IVC among many others. These anomalous pathways have various clinical implications and therefore need to be recognised and not be misguided incorrectly for pathology. The azygous vein accommodates blood from the lower half of the body and is thus dilated. It appears on the chest x-ray as widening of the mediastinum[6] and misinterpreted as para-vertebral or mediastinal mass. It can be rightly diagnosed using a computed tomography scan or a magnetic resonance imaging[7]. Moreover, IVC oddities predispose to augmented incidence of deep venous thrombosis which is usually caused by increased back pressure and venous stasis[2,6]. Elective procedures like right heart catheterization, transvenous device closure, IVC filter placements, electrophysiological studies, cardiopulmonary bypass surgery or any procedure requiring vascular access through femoral vein can be troublesome and may result in complications[2,6]. We aim to discuss the cardiac catheterization performed and the difficulties faced in described cases with interrupted IVC over a period of 2 years at the Armed forces institute of cardiology.

METHODOLOGY

This retrospective study was carried out at the Armed forces institute of cardiology/National institute of heart diseases Pakistan (AFIC-NIHD), and included 34 consecutive patients with interrupted IVC undergoing cardiac catheterization (both diagnostic as well as therapeutic) from January 2017 to December 2018. Data was collected after reviewing clinical records, catheterization and echocardiography reports and follow up records. Before undertaking catheter procedures, a detailed history, physical examination, complete blood counts, chest X-ray and a detailed transthoracic echocardiography was performed. Interrupted IVC was diagnosed on echocardiography in subcostal views and its anomalous drainage pattern was determined.

Cardiac catheterization procedures were effectuated as per our set protocols. Vascular accesses obtained were already decided on basis of anatomy and nature of study/procedure undertaken. The general approaches in diagnostic procedures were: 1) Femoral vein to azygous or hemiazygous vein to SVC to Right atrium to right ventricle to great artery/arteries depending upon underlying anatomy, 2) from internal jugular vein to desired chambers, 3) arterial approach especially in older children with Tetralogy of Fallot like anatomy. Details of catheterization procedure and problems encountered were recorded (table). Post procedural care followed standard post catheter protocol. The data was entered in SPSS 23 and descriptive analysis was done.

RESULTS

This study included 34 consecutive patients with interrupted IVC who underwent cardiac catheterization procedures [28 diagnostic procedures (82%) and 6 transcatheter interventional procedures] with mean age of 4.3 ± 4.5 years, including 19 males & 15 females. Age of study population ranged from 03 months to twenty years and included four infants aged 3, 7, 8 & 11 months. Mean height and weight was 95.5 ± 31 cms & 15 ± 11 kgs respectively. The mean amount of contrast used was 50 ml. The mean procedural time and fluoroscopy time were 55 & 17 minutes respectively and there were no major or minor complications. Three patients experienced transient arrhythmias but all recovered well. 88% cases were done under general anaesthesia with a 100% procedural success rate. Ninteen patients required both arterial and venous access, 12 required only a venous access and three patients needed only arterial access to complete the catheter procedure. From venous access perspective, 21 patients had only femoral access, 6 had internal Jugular venous access and three patients needed both femoral vein as well as internal

jugular vein to complete the study. No significant vascular complications were encountered.

The details of cases are shown in table. Among 28 diagnostic cardiac catheterizations, 20.6% were patients with underlying diagnosis of Tetralogy of Fallot (TOF) or similar anatomy including pulmonary atresia (PA), 17.6% were double outlet ventricle (DORV), 8.8% were unbalanced atrio-ventricular septal defects (AVSD), 8.8% transposition or corrected transposition of great arteries (TGA), 5.9% ventricular septal defects (VSD) with pulmonary hypertension (PHT), 5.9% partial AVSD & PHT, 5.9% partial anomalous pulmonary venous return (PAPVR), 2.9% atrial septal defect (ASD), 2.9% VSD & pulmonary stenosis (PS) and 2.9% VSD & PA Band. There were six transcatheter interventions encompassing 4 cases of Patent ductusarteriosus (PDA) device closure, one ASD device closure and one case of Major Aorto Pulmonary Collateral (MAPCA) coil occlusion.

Table: Details of procedures (n=34).

	Total (n)	Percentage (%)
Diagnostic procedures		
Tetralogy of Fallot / PA	07	20.6
DORV, TGA, VSD, PS	06	17.6
Unbalanced AVSD, PS	03	8.8
CTGA or TGA, VSD, PS	03	8.8
VSD, PHT	03	8.8
Partial AVSD, PHT	02	5.9
PAPVR, ASD	02	5.9
VSDs, PA Band	01	2.9
VSD, PS	01	2.9
Interventional procedures		
PDA Device Closure	04	11.8
ASD Device Closure	01	2.9
MAPCAs coil occlusion	01	2.9
Total	34	100

ASD device closure was done for a 16-year-old girl with 24 mm ASD secundum with interrupted IVC and left SVC to coronary sinus requiring a right internal Jugular venous approach. A 27 mm ASD device was deployed successfully under transesophageal echocardiography guidance. Four PDA device closures were done in patients with Interrupted IVC; two were done from right internal jugular vein and two from right femoral venous route through azygous vein to right atrium to right ventricle to pulmonary artery and to PDA to aorta. One 5-year-old boy was successfully offered PDA device closure through arterial line as he required smaller device and delivery sheath.

Complications like death, non-fatal cardiac arrest, life threatening arrhythmias, heart perforation or cardiac tamponade, bleeding/hematoma or vascular complications were not encountered in our study population.

DISCUSSION

Interrupted IVC is an uncommon vascular anomaly in general population and results from failure of fusion of the hepatic and renal segments of the inferior vena cava resulting in blood drainage to SVC and right atrium through the azygous or hemiazygous veins[8]. The hepatic veins drains directly in the right atrium. The procedures like right heart catheterization, transvenous device closure, IVC filter placements, electrophysiological studies, cardiopulmonary bypass surgery or any procedure requiring vascular access through femoral vein can be troublesome and may result in grave complications[2,6]. Consequently, it is important to know the exact anatomy before these procedures to avoid on table surprises[9]. Our experience of 34 cardiac catheterization /procedures in a relatively small cohort of young patients (including four infants) is presented. The most common diagnostic procedure was diagnostic cardiac catheterization for TOF in seven patients; three of them needed both arterial and femoral vein lines, two required only femoral vein approach and two catheter studies were completed by taking only femoral arterial access. In comparison, only femoral venous access is required to complete study in 75% of TOF patients with normal vascular anatomy as per our institution experience. Changela *et al* reported incidence of 0.5% of interrupted IVC in 2235 studied patients with TOF patients[10]. It is also worth mentioning that catheter study is challenging in setting of single ventricle physiology with severe PS & interrupted IVC, especially

while approaching the branch pulmonary arteries through internal jugular vein. In these patients, chiefly of tricuspid atresia with small VSD, an arterial approach should be initially attempted.

In our study, there are 6 transcatheter interventions including 04 PDA device closure, 01 ASD device closure and one MAPCA coil occlusion. Both ASD & PDA device closure in patients with interrupted IVC is an arduous task as it involves difficult routes, kinking of delivery sheaths, possibility of arrhythmias and hemodynamic compromise. For ASD device closure the hepatic venous approach could be an attractive alternative as it offers straighter path to left atrium and is being widely reported in literature. We performed an ASD device closure in a young girl choosing a right internal jugular venous approach but it took a while to cross the ASD. The ASD device was then deployed while keeping the wire still in left atrium. Truong *et al* recently reported their experience of successful ASD device closure using femoral venous & internal Jugular venous approaches[9,11]. Yucel *et al* reported use of steerable catheters in ASD device closure[12]. Having said that, PDA device closure in patients with interrupted IVC can also be done using various approaches[13-17]. We also executed 4 PDA device closures; from right internal Jugular vein, right femoral venous route and through arterial approach. Apart from kinking of the delivery sheath, there was no other problem even from femoral venous approach[18].

The mean procedural time of 17 minutes in our study was double than expected with similar catheter studies in patients without interrupted IVC. Three patients experienced transient arrhythmias but all recovered well. There were no fatal complications encountered in study population.

CONCLUSION

Cardiac catheterizations in patients with interrupted IVC can be challenging especially while carrying out device closure of ASD and PDA. Pre-procedural assessment is of paramount importance to plan an ideal and most favourable approach thus avoiding on table surprises.

CONFLICT OF INTEREST

This study has no conflict of interest to be declared by any author.

REFERENCES

1. Al Mandhari H, Rolnitsky A. Interrupted inferior vena cava with azygos continuation in an extreme preterm infant: Unusual lower-limb peripherally inserted central catheter line tip position. J Clin Neonatol 2019; 8(1): 122-4.
2. Goyal BK, Lahoti HA. Interrupted Inferior Vena Cava with Hemiazygous Continuation Causing Difficulty in Right Heart Access. J Heart Stroke 2016; 1(3): 1012-15.
3. Bass JE, Redwine MD, Kramer LA, Huynh PT, Harris JH. Spec-trum of congenital anomalies of the inferior vena cava: Cross-sectional imaging findings. Radiographics 2000; 20(3): 639-52.
4. Ugas CCF, Alpaca RLR, Matos RIA, Lazarte RCI, Valdez QM, Katekaru TDA et al. Characterisation of computed tomography angiography findings in paediatric patients with heterotaxy. Pediatr Radiol 2019; 49(1): 1142-51.
5. Moore KL, Persaud TVN: Clinically Oriented Embryology. 8th ed. 2007: 536.
6. Mandato Y, Pecoraro C, Gagliardi G, Tecame M. Azygos and hemiazygos continuation: An occasional finding in emergency department. Radiol Case Rep 2019; 14(9): 1063-68.
7. Bartram U, Fischer G, Kramer HH. Congenitally interrupted inferior vena cava without other features of the heterotaxy syndrome: report of five cases and characterization of a rare entity. Pediatr Dev Pathol 2008; 11(4): 266-73.
8. Punn R, Olson I. Anomalies associated with a prominent azygos vein on echocardiography in the pediatric population. J Am Soc Echocardiogr 2010; 23(3): 282-85.
9. Truong QB, Dao AQ, Do NT, Le MK. Percutaneous atrial septal defect closure through femoral and transjugular approaches in patients with interrupted inferior vena cava. J Cardiol Cases 2018; 18(3): 106-9.
10. Changela V, John C, Maheshwari S. Unusual cardiac asso-ciations with Tetralogy of Fallot-a descriptive study. Pediatr Cardiol 2010; 31(6): 785-91.
11. Lowry AW1, Pignatelli RH, Justino H. Percutaneous atrial septal defect closure in a child with interrupted inferior vena cava: successful femoral venous approach. Catheter Cardiovasc Interv 2011; 78(4): 590-93.
12. Yücel İK, Ballı Ş, Küçük M, Çelebi A. Use of steerable delivery catheter to successfully deliver a Ceraflexseptaloccluder to close an atrial septal defect in a child with interrupted inferior vena cava with azygos continuation. Turk Kardiyol Dern Ars 2016; 44(3): 244-47.
13. Akhtar S, Samad SM, Atiq M. Transcatheter closure of a patent ductusarteriosus in a patient with an anomalous inferior vena cava. Pediatr Cardiol 2010; 31(1): 1093-95.
14. Fernandes P, Assaidi A, Baruteau AE, Fraisse A. Transcatheter closure of a large patent ductusarteriosus using jugular access in an infant. J Thorac Dis 2018; 10(3): e207-e09.
15. Tefera E, Bermudez-Cañete R. Percutaneous closure of patent ductus arteriosus in interrupted inferior caval vein through femoral vein approach. Ann Pediatr Cardiol 2014; 7(1): 55-57.
16. Aggarwal N, Agarwal M, Joshi R, Joshi RK. Device closure of patent ductus arteriosus in interrupted inferior vena cava. Indian Heart J 2015; 67 Suppl 3: S85-S87.
17. Patel NH, Madan TH, Panchal AM, Thakkar BM. Percutaneous closure of patent ductus arteriosus via internal jugular vein in patient with interrupted inferior vena cava. Ann Pediatr Cardiol 2009; 2(2): 162-64.
18. Charlagorla P, Breinholt JP. Creating a Fontan fenestration in a child with dextrocardia and interrupted inferior vena cava. Ann Pediatr Cardiol 2016; 9(2): 179-82.