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ORIGINAL ARTICLE

Evaluating Types of TAPVR on Echocardiography and CT Angiography in Paediatric Patients Admitted in Tertiary Care Hospital: A Comparative Study

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ABSTRACT

Objective: To evaluate the types of TAPVR on Echocardiography and CT Angiography in Paediatric Patients

Study design: A comparative study

Place and Duration: Pediatric cardiology department and CT Angiography department of the National Institute of Cardiovascular Diseases (NICVD) Karachi from January 2019 to December 2021

Methodology: Patients who have TAPVR confirmed by CT angiography and detected by echocardiography were included in the research. Cases having anomalies in the systemic veins were eliminated. Demographic variables (age, gender, weight, etc.), echocardiographic results (such as type of TAPVR, obstructive and non-obstructive), and CT angiography findings were collected for the study using a preset structured proforma. We evaluated the diagnostic disparity between the two modalities.

Results: In the present study there were 107 patients, 67.3% were male gender, 56.1% with age less than 6 months, the median age was 6 months, median weight was 4.5 kg. On echocardiography, there were 83.2% non-obstructive, 47.7% supracardiac and 36.4% cardiac TAPVR. On CT angiography, there were 66.4% were non-obstructive, 53.3% supracardiac and 23.4% cardiac TAPVR. Of the associated cardiac diseases, in common 86.9% were found with atrial septal defect, 13.1% were persistent left superior vena cava, 12.1% were patent ductus arteriosus, 8.4% were a ventricular septal defect, 6.5% were of pulmonary atresia, and 3.7% were complete endocardial cushion defect.

Conclusion: As compared to echocardiography, CT angiography is more reliable for the detection of obstructive and mixed types of TAPVR. Detection of the supracardiac type of TAPVR is also almost the same as on echocardiography and CT angiography. As compared to CT angiography, echocardiography over diagnoses the non-obstructive type of TAPVR. CT angiogram is also useful in detecting other associated lesions such as heterotaxy, abnormal coronaries, presence of bilateral superior vena cava and coronary sinus.

Keywords: TAVPR, echocardiography, CT angiography, cardiac disorders

INTRODUCTION

Total abnormal pulmonary venous return (TAPVR), the sixth most frequent cause of congenital cardiac disease, is a rare syndrome that affects 0.6 to 1.2 out of every 10,000 live newborns (CHD). [1,2] TAPVR results from the common pulmonary veins' failure to connect to the left atrium due to the maintenance of the pulmonary veins' primitive connections with the cardinal systemic veins. [3, 4]

In this circumstance, saturated blood leaving the lungs and returning to the body takes an unusual route, returning to the systemic venous circulation rather than the left atrium. According to the location of the link, Darling et al. divided the lesions into four types: Cardiac TAPVC involves connections to the coronary sinus or right atrium; supracardiac TAPVC involves connections to the right superior vena cava, azygous vein, or left innominate vein [3] Mixed TAPVC combines any of the aforementioned connections and with Infracardiac TAPVC, which involves connections to the portal venous system, ductus venosus, or inferior vena cava below the diaphragm. The point of return to the systemic veins may be supracardiac, cardiac (coronary sinus or directly into the right atrium), Infracardiac, or both (superior vena cava and one of its draining veins) (inferior vena cava or portal system and often associated with obstruction). [5,6]

Although echocardiography is a highly effective diagnostic tool, there can be difficulties with identifying blockage, cases of mixed or Infracardiac variety, or TAPVR linked to heterotaxy syndrome or right atrial isomerism. Additional issues limiting reliable echocardiographic diagnosis in newborns with respiratory distress, particularly those on high-frequency oscillation mode of ventilation, include low acoustic windows, spatial resolution, and the operator's subjective interpretation. In complex situations, supplementary techniques like cardiac catheterization, cardiac CT angiography, or cardiac magnetic resonance imaging (cardiac MRI) may offer complete information. [7]

The 64-slice spiral CT angiography (64-SSCTA) procedure is frequently used to examine cardiovascular illness since it is non-invasive, quick to scan, has a large field of view, has the excellent spatial resolution, and has robust post-processing benefits [8]. The structure of the heart's anatomy and the arteriovenous connection can be demonstrated well using a variety of reliable post-processing images, which is crucial for the preoperative evaluation of infants with severe congenital heart disease [9]. To create a strong preoperative plan, an accurate diagnosis of TAPVR is essential. The purpose of this research was to present a clear concept for a reliable preoperative clinical diagnosis of TAPVR with an emphasis on the use of 64-SSCTA. [8, 9]

The anatomical form of TAPVR and the existence or absence of restriction to the drainage of these aberrant veins determine the surgical results. To provide a full diagnosis, it is crucial to identify the connections between all of the pulmonary veins, the drainage of the confluence, the vertical vein's path, and the presence of obstruction. [11, 12]

The surgical treatment of children born with a complete anomalous pulmonary venous connection (TAPVC) has significantly improved during the past ten years. In terms of lower morbidity and mortality as well as overall outcome, this has been reflected in the reported results from numerous centres. TAPVC is an uncommon defect that affects just 1.5% of children with congenital heart disease, although it frequently manifests early and requires neonatal urgent cardiac surgery. The results of TAPVC

have gotten better over time and reported death rates have consistently been under 10%. But the most challenging grouping appears to be mixed-type TAPVC, which has more than one level of pulmonary venous drainage. Patients with mixed TAPVC continue to die at significant rates in some series. [13]

It can occasionally be linked to heterotaxy syndrome, which has a high death and morbidity rate. The surgical repair of TAPVC remains difficult, even in cases of a biventricular architecture, despite recent breakthroughs in surgical procedures and postoperative care. [14, 15]

The purpose of the study is to evaluate the diagnostic efficacy of transthoracic echocardiography and CT angiography in the evaluation of different forms of TAPVC. This study compares and assesses the advantages of transthoracic echocardiography against CT angiography.

METHODOLOGY

This retrospective comparative study was conducted at the pediatric cardiology department and CT Angiography department of the National Institute of Cardiovascular Diseases (NICVD) Karachi from January 2019 to December 2021. Data for 2 years were collected from the record room of the department. Patients of any age diagnosed with TAPVR on echocardiography and diagnosis confirmed by CT angiography done at NICVD were included in the study. Patients with systemic venous abnormalities were excluded.

After receiving approval from the institutional ethical review committee, the National Institute of Cardiovascular Disease (NICVD) Hospital's Paediatric Cardiology Department extract the necessary study data from the patient records of TAPVR patients' ECHO and CT angiography departments for the duration of the study, which runs from January 2019 to December 2021.

The study identifies and includes patients whose TAPVR diagnosis was made by echocardiography and confirmed by CT angiography. Cases having anomalies of the systemic veins were disqualified. Demographic information (age, gender, weight, etc.), echocardiographic findings (such as kind of TAPVR, obstructive and non-obstructive), and CT angiographic findings were collected for the study using a preset structured proforma. We evaluate the diagnostic disparity between the two modalities. The necessary data is inserted on the sheet from the patient's hospital record database. Inputting the hospital registration number prevents data duplication.

Data were stored and analyzed using IBM-SPSS version 23.0; Counts with percentages were reported on gender, age group, and weight category. Median with Interquartile range was given on age and weight. Descriptive were reported on the type of TAPVR on echocardiography, CT angiograph and associated cardiac disease of studied samples.

RESULTS

In the present study there were 107 patients, 67.3% were male gender, 56.1% with age less than 6 months, the median age was 6 months with 25^{th} – 75^{th} percentile from 2.5 – 11 months, median weight was 4.5 kg with 25^{th} – 75^{th} percentile from 3.3 – 6.0 kg, on average 62.6% children were found with 2 – 5 kg weight. (As shown in Table 1)

Age at time of diagnosis on echocardiography was less than or equal to 6 months of 62.6% children, median was 5.0 months with 25^{th} – 75^{th} percentile from 1.5 – 10 months, median age at time of diagnosis of disease on CT Angiography was 5 months with 25^{th} – 75^{th} percentile from 2.0 – 9.0 months, on average 56.1% children were found with age less than or equal to 6 months. (As shown in Table 2)

The type of TAPVR on echo, there were 83.2% non-obstructive, 47.7% supracardiac and 36.4% cardiac TAPVR. There were 6 (5.6%) cases of obstructive type, while the Infracardiac types were 4 (3.7%) and mixed types were 6 (5.6%) (As shown in Table 3). The type of TAPVR on CT, there were 66.4% non-obstructive, 53.3% supracardiac and 23.4% cardiac TAPVR. There were 22 (20.6%) cases of obstructive type, 4 (3.7%) of Infracardiac type and mixed types16 (15%) (As shown in Table 4)

Regarding the associated cardiac diseases, in common 86.9% were found with atrial septal defect, 13.1% were persistent left superior vena cava, 12.1% were patent ductus arteriosus, 8.4% were ventricular septal defect, 6.5% were pulmonary atresia, 3.7% were complete endocardial cushion defect as associated cardiac disease. (As shown in Table 5)

Table 1: Baseline Characteristics of Studied Samples (n=107)

Characteristics		n	%
Gender	Male	72	67.3
	Female	35	32.7
Age group	≤6 months	60	56.1
	7 - 12 months	23	21.5
	>12 months	24	22.4
	Median (IQR)	6	2.5- 11
Weight (kg)	2 - 5 kg	67	62.6
	6 - 10 kg	26	24.3
	>10 kg	14	13.1
	Median (IQR)	4.5	3.3 – 6.0

Table 2: Age Distribution at diagnosis on ECHO and CT

Age Group at Echo and CT		n	%
Age at date of diagnosis on echo	≤6 months	67	62.6
	7 - 12 months	17	15.9
	>12 months	23	21.5
	Median (IQR)	5.0	1.5 – 10
Age at date of diagnosis of disease on CT Angiography	≤6 months	60	56.1
	7 - 12 months	22	20.6
	>12 months	25	23.4
	Median (IQR)	5.0	2.0 – 9.0

Table 3: Type of TAPVR on ECHO

Type of TAPVR on echo	n	%
Supracardiac	51	47.7
Non obstructive	89	83.2
Obstructive	6	5.6
cardiac	39	36.4
Infracardiac	4	3.7
Mixed	6	5.6

Table 4: Type of TAPVR on CT Angiography

Type of TAPVR on CT Angiography	n	%
Supracardiac	57	53.3
Obstructive	22	20.6
Non-obstructive	71	66.4
cardiac	25	23.4
Mixed	16	15
Infracardiac	4	3.7

Table 5: Associated cardiac disease

Associated cardiac disease	n	%
Atrial septal defect	93	86.9
Patent ductus arteriosus	13	12.1
Persistent left superior vena cava	14	13.1
Ventricular septal defect	9	8.4
Heterotaxia syndrome with polysplenia	4	3.7
Osteal LPA stenosis	2	1.9
Pulmonary atresia	7	6.5
Complete endocardial cushion defect	4	3.7
Single ventricle	1	0.9
Transposition of the great arteries	2	1.9
COA	2	1.9
Pulmonary valve under stenosis	2	1.9
Svc aneurysm	1	0.9
Patent ductus arteriosus	1	0.9
TOF	1	0.9
Patent foramen ovale	2	1.9
Single ventricle	1	0.9
Dextrocardia	2	1.9

In our study there were 5.6% were found with incidental findings on CT scan, 0.9% were situs ambiguous on PAPVR on echo, on PAPVR on CT angiography 1.9% were right-sided veins opening into RA, 0.9% were right-sided veins opening in SVC, and 0.9% were left-sided veins into left-sided RA.

DISCUSSION

In the present study, there were 107 patients , 67.3% were male gender, and 56.1% with age less than 6 months. A similar local study included 47 patients in total, of whom 33 (70%) were males. The median age was three months, with ages ranging from one day to 168 months. Weights varied from 1.8 kg to 39 kg, with 3.5 kg serving as the median. In 24 patients and 30 patients, the results of cardiac CT angiography and echocardiography were compared. Prior to surgery, cardiac CT angiography was conducted on seven patients, and the results were similar to what was noted in the surgical notes. [7] While other results were consistent with CT angiography or surgical diagnosis, echocardiography missed the identification of two related congenital cardiac anomalies: one case of persistent left superior vena cava and one case of pulmonary atresia. The diagnostic value of echocardiography was shown to be modest (27%) in the diagnosis of mixed variety and TAPVR combined with other CHD, but very sensitive (81%) in the diagnosis of isolated TAPVR. [7] In another study TAPVR (Total anomalous pulmonary venous return) and PAPVR (Partial anomalous pulmonary venous return) types of pulmonary venous abnormalities were accurately represented by MDCT with 100% sensitivity and 100% specificity. [16] Dalia F Elbeih et al concluded that the concordance between CTA findings and echocardiographic findings for SVC anomalies, IVC anomalies, and pulmonary venous anomalies was 77.8%, 66.7%, and 90%, respectively. [17]

In this study type of TAPVR on echo, there were 83.2% non-obstructive, 47.7% supracardiac and 36.4% cardiac TAPVR. On the type of TAPVR on CT, there were 66.4% non-obstructive, 53.3% supracardiac and 23.4% cardiac TAPVR. Tain Lee et al found that among 14 newborns in all, none of the newborns required any additional diagnostic cardiac catheterization. Only one atrial septal defect was missed in a patient with coarctation syndrome, resulting in a 98% (53/54) accuracy rate for diagnosing distinct cardiovascular defects with MDCT. [18] In an international study transthoracic echocardiography had a sensitivity of 86.5% and a specificity of 100% for total anomalous pulmonary venous connection (TAPVC). Transthoracic echocardiography has a 66.6% sensitivity and 100% specificity for isolated pulmonary vein stenosis. On transthoracic echocardiography, they were unable to evaluate pulmonary venous drainage in 38 (38.5%) of the patients. [19] A case series study outline two instances when echocardiography was inconclusive. Instead of cardiac catheterization, a contrast-enhanced computerized tomography (CT) scan of the chest and abdomen was carried out to make the diagnosis. When echocardiography is ambiguous, they propose that CT with contrast is a noninvasive technique to gain anatomic details of pulmonary venous drainage in TAPVR. [1]

Our results found associated cardiac diseases, in common 86.9% were found with atrial septal defect, 13.1% were persistent left superior vena cava, 12.1% were patent ductus arteriosus, 8.4% were a ventricular septal defect, 6.5% were pulmonary atresia, 3.7% were complete endocardial cushion defect as associated cardiac disease.

In our study 6 cases of supracardiac TAPVR were missed on echocardiography that was later on detected on CT angiogram, while 16 cases of obstructive types were missed on echocardiography. A total of 89 cases were diagnosed as a non-obstructive type of TAPVR on echocardiography, while on CT angiogram only 71 cases were confirmed as non-obstructive TAPVR.

The detection rate of the Infracardiac type of TAPVR was the same on echocardiography as well as CT angiogram. Regarding the mixed type of TAPVR 16 cases were missed on echocardiography. As echocardiographic findings are operator-dependent, so we expect this kind of result necessitates the need for a CT angiogram.

CONCLUSION

As compared to echocardiography, CT angiography is more reliable for the detection of mixed types of TAPVR. Detection of the supracardiac type of TAPVR is also almost the same as on echocardiography and CT angiography. As compared to CT angiography, echocardiography over diagnoses the non-obstructive type of TAPVR. CT angiogram is also useful in detecting other associated lesions such as heterotaxy, abnormal coronaries, presence of bilateral superior vena cava and coronary sinus.

Recommendations: This was a pilot project, that we did in our setup to see the prevalence of TAPVR and its various types. Further studies are suggested in which these patients are followed up for the management offered to them and the chances of survival, prognosis after surgery and quality of life.

Further studies are needed to check these things which will tell us the likelihood of survival of such patients in our setup. Usually in our observation patients presented late and already pulmonary hypertension has developed, so patients could be operated but because of severe chest infections they are not fit for surgery. So we suggest fetal echocardiography on the patients suspected of congenital heart disease especially TAPVR, as early diagnosis could be made or at least neonatal screening should be done on these patients so that early diagnosis and management could be done. Furthermore, Infracardiac TAPVR patients and obstructive TAPVR patients have a very poor prognosis in our setup due to the nonavailability of neonatal surgical expertise in many cities.

In case of difficult TAPVR cases, like mixed type, obstructive type, Infracardiac type, complex congenital heart disease and associated anomalies CT angiogram is recommended apart from screening echocardiography.

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