MAR Cardiology & Heart Diseases (2023) 3:8

Research Article

Is Routine Cardiac Catheterization Necessary before Cardiac Surgery in Children with Tetralogy of Fallot (tof)?

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Received: 13 December 2023

Published: 30 December 2023

Abstract

Although echocardiography is well established as the first-line imaging technique for the diagnosis of all forms of congenital heart disease, most institutions continue to perform cardiac catheterization prior to surgical repair of more complex defects such as TOF. In this study we want to determine the diagnostic value of echocardiography compared with cardiac catheterization in children with TOF.

To determine the diagnostic value of echocardiography compared with cardiac catheterization in children with TOF, we reviewed the records of 99 children with TOF at Imam Reza Hospital of Mashhad. We included children with TOF who underwent echocardiography plus cardiac catheterization prior to surgical repair. We defined "major differences" as those that effect on surgical plan and "minor differences" as those that did not. Also we defined "false further finding" as those that determined only by echocardiography.

***Results**: From 99 patients 55 were male, 82% had normal birth. The mean age of diagnosis and surgery was 1.55 and 5.42yrs respectively. Only 4 percent (4 of 99) had major differences such as: Coronary artery anomalies (2 cases), small PA branch plus MAPCAs (1), absence of LPA or RPA (1). Fifty-six cases had minor differences: 28% PA branch origin stenosis (28 of 99), ASD (15%), right aortic arch (7%), LSVC (4%), PDA (2%), small PA branch (2%), muscular VSD (2%), RV dysfunction (2%), aberrant RSCA (1%), abnormal arborization (1%). Forty cases had no differences. Twenty-three percent had false further diagnosis and the most common was small PA branch (6%). operation type in78% was complete repair (46 of 59) and mortality rate was 3%, complication was seen 54.8% and 22% had excellent results.*

***Conclusion**: This study suggests that echocardiography alone is an accurate tool for the preoperative diagnosis of TOF in most (96%) children undergoing surgical repair esp. in conjunction of alternative noninvasive methods such as cardiac magnetic resonance imaging and CT angiography and may obviate the need for routine diagnostic catheterization.*

***Keyword**: Echocardiography, Cardiac catheterization, Children, Tetralogy of Fallot, Diagnostic value, Congenital heart disease.*

Introduction

Congenital heart disease is currently a major public health problem in all countries, especially in industrialized countries (1). Despite significant advances in the treatment of these diseases over the past years, heart defects make up a large proportion of the causes of neonatal mortality. In addition, many Patients who have had surgery will not receive definitive treatment and will face further problems and mortality, tetralogy of Fallot (TOF) is one of the most common cyanotic congenital diseases. In this study, we will compare these two common diagnostic methods in pre- op evaluation before surgery (2,3)

Diagnosis

As with most diseases, there is a wide range of heart disease severity. Sever diseases are easy to diagnose. Each of the severe defects has obvious symptoms. Also may lead to abnormal heart function, The diagnosis of mild forms may be delayed (2).

In a chest radiography (CXR), the typical anterior-posterior view includes the narrow base of the heart, the concavity of the left border of the heart in the area usually occupied by the pulmonary artery, and the normal size of the heart. Hypertrophy of the right ventricle causes the shadow of the tip of the heart to rotate upward. In this case, the tip of the heart is more than normal above the diaphragm. The heart shadow is thought to be like a boot or shoe. The areas of the umbilical cord and lungs are clear and clean because of reduced pulmonary blood flow or the small size of the pulmonary arteries, or both. The aorta is usually large and in 20% of cases towards It bends to the right, causing the toothed shadow of the trachea and bronchi to jag in the anterior-posterior view and its deviation to the left (3).

The Electrocardiogram (ECG) shows right-axis deviation and evidence of right ventricular hypertrophy. A clear and long R is seen in the right anterior heart derivatives. In some cases, the only sign of right ventricular hypertrophy is initially a positive T-wave in V3R or V 1 Leads. The P-wave is sharp and sometimes bipolar (3).

Two-dimensional (2D) echocardiography confirms the diagnosis and provides information about the extent of aortic mounting on the interstitial wall, the location and severity of right ventricular outflow obstruction, the size of the pulmonary artery branches, and the orientation of the aortic arch.

Echocardiography is also useful in determining whether the regenerative duct supplies part of the blood flow to the lungs. Echocardiography may eliminate the need for cardiac catheterization (3).

Cardiac catheterization shows that right ventricular systolic pressure is equal to systemic pressure. If it enters the pulmonary artery, the pressure drops dramatically, although crossing the right ventricular outflow tract, especially in severe cases, can cause a hypoxic attack. Pulmonary artery pressure is usually 10 to 15 mm Hg lower than normal. The level of arterial oxygen saturation strongly depends on the right shunt to the left. In patients with resting cyanosis, this value is about 75 to 85% (3).

Selective right ventriculography best shows the anatomy of the tetralogy of the fallot. Infundibular stenosis can vary in length, width, shape, and elasticity. The pulmonary valve is usually thickened and the annulus may be small. In patients with pulmonary atresia and ventricular septal defect, the anatomy of the pulmonary arteries may be quite complex and, for example, there may be no connection between the right and left pulmonary arteries. Complete and accurate information about the anatomy of the pulmonary arteries is important at the time of evaluation of these patients who are candidates for surgery (3).

Left ventriculography shows the size of the left ventricle, the position of the ventricular wall defect, and the aorta mounted on it. It also proves the connection between the mitral and the aorta and can therefore reject the right ventricle with two outlets. Coronary aortography or arteriography determines the path of the coronary arteries. In 5 to 10% of patients with tetralogy of the phallus, a large misplaced coronary artery passes through the right ventricular outlet. This artery should not be cut during surgical repair. Ensuring that the anatomy of the coronary arteries is normal is important in planning surgery for young children who may need to have a patch implanted in the pulmonary valve. Echocardiography can determine the anatomy of the coronary artery, and angiography is performed only in cases that are Complex (3).

Prognosis and complications

Prior to correction, patients with follow-up tetralogy are prone to several serious complications. Fortunately, most children experience Partial or complete repair during childhood, and these side effects are rare. Cerebral thrombosis is common in the cerebral veins and sometimes in the cerebral arteries,

Brain abscesses are less common than cerebrovascular complications and are very rare when many patients recover at a younger age, Infectious endocarditis may occur, Heart failure is not common in patients with tetralogy. (3).

Accompanying anomalies

The Patent ductus arteriosus (PDA) may be present and sometimes atrial septal defects (ASD) are seen. The right aortic arch is seen in 20% of patients with tetralogy of Fallot, and other abnormalities of the pulmonary arteries and aortic arch may also be seen. Persistence of the left superior vena cava (LSVC) and its drainage into the coronary sinus may be observed. Sometimes there are multiple ventricular septal defects(VSD) and they must be diagnosed before corrective surgery (3). The absence of a pulmonary artery branch, which in most cases is a left branch, should be suspected if the radiographic appearance of the pulmonary arteries differs on both sides. Absence of a pulmonary artery is often associated with hypoplasia of the affected lung. The absence of a branch of the pulmonary artery must be diagnosed because obstruction of the remaining pulmonary artery during surgery severely reduces pulmonary blood flow, which is already reduced (3).

Methods

Patient population

This cross-sectional descriptive study was performed on 99 patients with tetralogy of Fallot who were evaluated and underwent echocardiography and angiography in Imam Reza Hospital during 5 years.

Inclusion and exclusion criteria

Patients under 18 years of age, pediatric population with Extreme TOF, TOF with pulmonary atresia and tetralogy of Fallot without any other complex heart defect who were evaluated and underwent both echocardiography and angiography.

Data collection

The sampling method was non-probabilistic in which the records of all patients with TOF during this period were reviewed.

Data collection tools in this study were an interview form and an observation form, an interview form to collect demographic data including year of birth, birth weight, referral weight, sex, age of diagnosis, age of first visit to the clinic, age of surgery. The form of observation includes echocardiographic and angiographic results, type of heart abnormality, type of operation and the result of existing surgery and patients.

Statistical analyses

Data were collected and processed through descriptive and inferential statistics and analyzed. In the descriptive statistics section, the absolute and relative frequency distributions as well as the means and scatter indices were used to describe the population. In inferential statistics, independent t-test, chi-square was used and correlation coefficient used between these two diagnostic methods. catheterism and angiography were considered as standard gold, then cases that have been diagnosed in echo and under diagnosis or over diagnosis, as well as sensitivity and specificity in general and separately were Calculated.

Results

Ninety-nine patients with TOF were studied. Forty-four patients (44.4%) were female and 55 patients (55.6%) were male. The patients' median age was 4.73 yrs. (from birth days to 18 years of age); 81% of the subjects were in the normal range for birth weight, which is expected according to the available information and sources about congenital heart disease. (1)

1.5% of patients had VLBW (less than 1500 g) and 15.2% of patients had LBW (1500 to 2500 g),1.5 %weighed more than normal. According to the age of the patients, 48.1% of the patients' weight was lower than the 5th percentile and were 2.5% higher than the 95th percentile, the mean age of surgery in patients is 5.425 yrs, the lowest of which was in infancy and the highest age was 18yrs. The most common age group was one to three years. The average delay in surgery in patients is 3.76 yrs, the

lowest of which was less than one month and the highest was 16.75 yrs. The most common delay in surgery was 1.66 to 3.3 yrs.

78 patients (89.7%), it was recommended to perform complete correction surgery and in 10.3% of cases, it was recommended to perform palliative surgery (shunt). 59 patients who underwent surgery, 78% were in complete correction surgery. And 22% of them have undergone palliative care. Parameters are summarized in (Table 1).

Table 1- Baseline characteristics in study patients

Variables	**Values**
Number of patients	99
Female	(44.4%)
Male	(55.6%)
Age, year	4.7 (0.1 - 18)
Mean age of surgery (year)	5.4 (1 - 18)
Delay in surgery (year)	3.7(0.1-16.75)
Recommended complete correction surgery	89.7%
Perform complete correction surgery	78%
weight lower than the 5th percentile	48.1%

Patients who have undergone surgery are divided into three categories according to the result of postoperative echo: The first group an acceptable postoperative echo result, second were complication group and the third group of patients who died during or after the operation.

The prevalence of complications based on postoperative echo include the following: Residual VSD (6.84%), Pleural effusion or pericardial effusion (25%). (Table 2).

Table 2: Distribution postoperative complications

Complications	Percentage
Residual VSD	6.84%
Pericardial and pleural effusion	25%
Arrythmia	9.6%
Moderate to severe residual PS	7.7%
Loss of consciousness	1.9%
Pneumothorax	1.9%
Diaphragmatic paralysis	1.9%

The most common types of minor differences between echocardiography and catheterization data in this study were: ASD (15.2%), Right aortic arch (7.1%), Small pulmonary artery branches (7.1%), LSVC (4).The types of major differences between echocardiography and catheterization data in this study were: separation of LAD from RCA (2%), smallness of pulmonary artery branches with MAPCAS (1%), Absent of a branch of Pulmonary artery (1%)

The finding that Echo reported in addition to angiography were classified as overdiagnosis "excess diagnosis"(22%). The types of this "excess diagnosis" were: Small pulmonary artery branches (7%), Stenosis in the origin of the branches of the pulmonary artery (5%), ASD or PFO (4%) (Table 3,4,5)

Table 3: Distribution of patients according to over diagnosis by echo

"Excess diagnosis" by Echo	Percent
Stenosis of the origin of the pulmonary artery branches	5
Right aortic arch	1
Dilatation of the coronary sinus	1
ASD or PFO	4
Small pulmonary artery branches	7
PDA or collateral	3

Aberrancy right Subclavian artery	1

Table 4: Evaluation of diagnostic power of echocardiography

Echocardiography Finding	Negative predictive value	positive predictive value	specificity	sensitivity
Right aortic arch	0.92	0.94	0.99	0.68
Branch origin stenosis	0.70	0.54	0.91	0.21
Patent foramen Oval	0.75	0.82	0.87	0.67
Persistent left superior vena cava	0.95	1.00	1.00	0.50
Small PA branch	0.91	0.43	0.95	0.27
Patent Ductus Arteriosus	0.97	0.83	0.93	0.93
Muscular VSD	0.98	1.00	1.00	0.33
Small LV	0.99	_	1.00	0.00
Right ventricular dysfunction	0.98	_	1.00	0.00
Aberrant RSCA	0.99	0.00	0.99	0.00
Abnormal arborization	0.99	_	1.00	0.00
Dilated CS	1.00	0.00	0.99	_
Major different	0.96	1.00	1.00	0.20

Table 5 : Difference between echo and angiography finding

variable	No	Minor	Minor+ Major	Major
N=99	40	55	2	2

Discussion

Echocardiography is A valuable tool in diagnosis of TOF and can define the VSD features, identify the degree of RVOT obstruction, and evaluate the general cardiac anatomy and function. Echocardiography data can help assess the patient's physiology and identify patients at higher risk for hyper cyanotic spells. Serial echocardiograms can give us many clues regarding the TOF physiology, but ultimately the categorization of "pink" vs "blue" tetralogy is a clinical diagnosis.

Echo is also used to guide surgical repair, with particular attention to VSD size and location, degree of RVOT obstruction, pulmonary valve and pulmonary artery size and stenosis, presence of coronary artery crossing the RVOT, and diagnosis of other associated lesions that may need to be addressed concurrently. In terms of VSD closure, differentiating between peri-membranous outlet vs. muscular outlet vs. doubly committed VSD can affect how the surgeon performs the repair and the suture lines chosen in order to avoid damage to the cardiac conduction system. (4,5)

The decisions about the complex diagnostic and therapeutic plans have to go through the analysis of all diagnostic techniques applied in these patients, as echocardiography, CMRI, CT, hemodynamic studies, electrophysiology, and biochemistry studies. Furthermore, the introduction of more quantitative echocardiographic parameters for the evaluation of the RV has not decreased the number of cardiac catheterizations but it has indeed improved the added value of these invasive procedures probably making more accurate the indications to perform them.(6)

In 95 cases out of 99 patients (96%) the diagnosis of echo alone and without the need for angiography is sufficient to guide the surgeon and only in 4 cases (4%) there was a need for additional study with angiography, In this study, with a significant difference, echo reduced the need for angiography and Help to Appropriate decision on the type and time of surgery.

Treatment of children with tetralogy of Fallot (TOF) has become more advanced in the last decades with increasing use of primary surgical repair and trans catheter right ventricular outflow tract palliation and fewer systemic-to-pulmonary shunts. With proper anatomy, there is a greater tendency for complete repair, in this study 78% were Treated as complete correction surgery and 22% of them have undergone palliative surgery. And decision-making based on echo results had no effect on the final choice of type of surgery for patients.

Patients who have undergone surgery are divided into three categories according to the result of postoperative echo. The first group of people who reported an acceptable postoperative echo result. The second as the "complication" group and the third group of patients who died during or after the operation. According to the studies performed, 21.7% of all patients who underwent surgery were acceptable, 54.84% had complications after surgery, and 2.9% died. Compared to the studies performed, it seems that the mortality rate of the operation was in the same range, but the postoperative complications in our study were higher than other cases

In terms of diagnostic differences between echocardiography and angiography:

In this study, echo diagnosis was compared with angiography and the types of defects and diagnostic differences of echo compared to angiography were divided into two categories of minor and major differences. According to this definition, minor difference refers to a difference that does not affect the surgeon and major refers to the difference that affects the surgeon operation. As can be seen, the sensitivity of echocardiography for the diagnosis of PDA (93%) and above all, followed by the sensitivity for the right aortic arch (68%) and PFO (67%).

The sensitivity of echocardiography is low in Items that change the surgeon Decision (major) (20%), so echocardiography is not a powerful tool to diagnose them, but the very low prevalence of these disorders in our study, as well as the potential risks of angiography (It is invasive and expensive), raising the question of whether all children with tetralogy should undergo angiography before surgery?

Conclusion

This study suggests that echocardiography alone is an accurate tool for the preoperative diagnosis of TOF in most children undergoing surgical repair and may obviate the need for routine diagnostic catheterization.

Acknowledgement

Thanks to the esteemed research assistant of the Mashhad University of Medical Sciences and the medical records department of Imam Reza Hospital.

The mentioned article is the result of a research project and a student thesis and has been approved by the ethics committee of Mashhad University of Medical Sciences.

Conflict of interest : there is no Conflict of interest.

Limitations

limited sample size and low incidence.

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