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Severe Residual Pulmonary Stenosis after Surgical Repair of Tetralogy of Fallot: What's Our Next Strategy?

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Abstract:- This case report describes a 25-year-old woman with Tetralogy of Fallot (TOF) who underwent surgical repair at age 15. Eight years later, she presented with recurrent chest pain and was found to have residual severe pulmonary stenosis and regurgitation. This is a complication of TOF repair, particularly when the pulmonary valve is not preserved. The paper discusses the management of TOF, including initial palliative procedures and complete repair surgery. It highlights the long-term complications such as pulmonary valve regurgitation and residual stenosis, both of which may require re-intervention. The importance of long-term follow-up for TOF patients is emphasized.

Keywords:- *Tetralogy of Fallot (TOF); Pulmonary Stenosis; Pulmonary Regurgitation; TOF Repair; Long-Term Complications; Single Ostium Coronary Transverse in RVOT.*

I. INTRODUCTION

Tetralogy of Fallot (TOF) is the predominant form of cyanotic congenital cardiac disease. The condition manifests in approximately 1 in every 3500 live births and represents 7-10% of all cases of congenital heart disease. The cause of TOF is complex, involving multiple factors, with 25% of cases being associated with chromosomal abnormalities. Isolated pulmonary stenosis (PS) occurs in around 4% to 8% of all cases of congenital heart abnormalities (CHDs). Pulmonary stenosis (PS) is frequently linked to other congenital heart defects (CHDs), including tetralogy of Fallot (TOF), single ventricle, and various others. Pulmonary stenosis (PS) can occur in the valve, below the valve (infundibular), above the valve (supravalvular), or within the right ventricular (RV) cavity (Park, et al 2020). The objective of TOF repair is to eliminate intracardiac shunting by closing the anterior malalignment ventricular septal defect and relieving RV outflow tract obstruction, while ideally preserving the pulmonary valve and protecting normal myocardial function. Therefore, surgery is the primary treatment option. The majority of patients with TOF do not show any symptoms during the newborn period and will have primary surgical treatment between the ages of 3 and 6 months. Individuals displaying symptoms have the option of receiving either primary repair or first palliation (either through surgery or transcatheter methods), followed by delayed repair, as suggested (Alboliras, et al 2018).

Severe residual pulmonary stenosis after surgical repair of Tetralogy of Fallot poses a complex challenge in patient management. The current surgical approach prioritizes the preservation of the pulmonary valve, even if it leads to mild residual stenosis (Latus et al., 2012). Patients who have undergone Tetralogy of Fallot repair may develop severe pulmonary regurgitation, necessitating either surgical or catheter-based valve replacement (Strozzi et al., 2019). Surgical repair of Tetralogy of Fallot can result in various residual findings, both early postoperatively and during long-term follow-up (Stambach & Valsangiacomo, 2009). Late complications following Tetralogy of Fallot repair include issues such as arrhythmias, ventricular dysfunction, and sudden deaths (Karamlou et al., 2006). The long-term outcomes of adult patients who have undergone surgical correction for Tetralogy of Fallot show a risk of residual lesions, valve regurgitation, ventricular dysfunction, arrhythmias, and sudden deaths (Haneda et al., 1990).

In managing severe residual pulmonary stenosis after surgical repair of Tetralogy of Fallot, considerations for pulmonary valve replacement may arise. Studies have emphasized the importance of timing and the outcomes of pulmonary valve replacement for pulmonary regurgitation in repaired Tetralogy of Fallot (Muhll, 2019). Additionally, preserving the pulmonary valve during Tetralogy of Fallot repair is a strategy that should be pursued to mitigate the risk of pulmonary regurgitation (Guariento et al., 2022). Moreover, the association between Down syndrome and pulmonary hypertension may contribute to more severe pulmonary regurgitation post-Tetralogy of Fallot repair, potentially necessitating earlier pulmonary valve replacement in these patients (Sullivan & Frommelt, 2017).

Dynamic obstruction of the right ventricular outflow tract can be seen in the immediate period following bypass with Residual right ventricular outflow tract obstruction post surgery is common but if in mild condition. Based on that in this article we report a case female with severe pulmonary stenosis following tetralogy of fallot repair. In conclusion, managing severe residual pulmonary stenosis after surgical repair of Tetralogy of Fallot necessitates a comprehensive understanding of the long-term implications

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of the repair, the development of late complications, and the potential need for pulmonary valve replacement to address issues such as pulmonary regurgitation. Close monitoring and timely interventions are crucial in optimizing outcomes for Tetralogy of Fallot patients.

II. LITERATURE REVIEW

Severe residual pulmonary stenosis after surgical repair of Tetralogy of Fallot poses significant challenges in the management of patients. Pulmonary valve regurgitation is a common residual issue post-repair, leading to right ventricular dilatation, dysfunction, heart failure symptoms, arrhythmias, and even sudden death (Buechel et al., 2005; Lee & Kwak, 2013). Chronic pulmonary regurgitation and residual pulmonary stenosis are among the chronic issues faced by individuals post-surgical repair, impacting long-term outcomes (Bailliard & Anderson, 2009). Early postoperative remodeling after repair typically involves significant pulmonary regurgitation, right ventricular hypertrophy, and lack of dilation (DiLorenzo et al., 2018).

Balloon pulmonary artery angioplasty and percutaneous balloon pulmonary valvuloplasty are interventions that may be considered in cases of severe vascular stenosis or residual pulmonary insufficiency, emphasizing the importance of lifelong follow-up to monitor these conditions (Warnes et al., 2008; Rao, 2007). Risk factors for severe pulmonary regurgitation post-repair include specific ratios and longer follow-up durations (Apandi et al., 2020). Surgical pulmonary valve replacement remains the gold standard for managing pulmonary regurgitation, even with advancements in interventional procedures (Mkalaluh et al., 2019).

Studies have highlighted the importance of timing pulmonary valve replacement based on specific criteria such as regurgitant fraction and anatomic or hemodynamic abnormalities indicating ventricular dysfunction (Mosca, 2016). Late pulmonary valve replacement has shown to improve right ventricular function, functional class, and reduce atrial arrhythmias with low mortality rates (Discigil et al., 2001). Residual pulmonary stenosis post-repair can lead to pressure overload on the right ventricle, compounding the existing volume overload (Tandia et al., 2022).

In conclusion, the management of severe residual pulmonary stenosis after surgical repair of Tetralogy of Fallot requires a comprehensive approach that considers the long-term implications of pulmonary valve regurgitation, the timing of interventions like pulmonary valve replacement, and the need for vigilant follow-up to address potential complications effectively. Surgical interventions remain crucial in addressing severe pulmonary regurgitation, emphasizing the importance of individualized care strategies tailored to each patient's specific needs.

III. CASE REPORT

Female patient 25 years od referred from Anuntaloko Parigi Regional Hospital to our Hospital with a diagnosis of Residual Pulmonary stenosis\ Severe post ToF repair in 2015. The patient was a referral from Harapan Kita National Hospital to underwent follow up post operation by Right Heart Catheterization. Currently complaining of left chest pain for the last 3 weeks. Previous history of pain since childhood. History of Bluish is known as a child aged 3 years, often blue when crying. There is no shortness of breath. There was previously a history of shortness of breath, the shortness of breath gets worse with heavy activity. The patient was the second of three siblings, born normal, immediately cried, in the hospital, birth weight 3500 grams and birth length not remembered. There was no history of maternal herbal consumption during pregnancy. There was no history of ARI during pregnancy A history of congenital heart disease was discovered at the age of 3 years There was no family history of congenital heart disease. On physical examination we found patent with compos mentis GCS E4V5M6, blood pressure: 123/104 mmHg, pulse: 75 x/minute regular, respiratory rate: 22 x/minute, axillary temperature: 36.7 °C and oxygen saturation around 97%-99% on all four extremities in room air . The conjunctiva not anemic, the sclera is not icteric, jugular vein pressure R+2 cmH20, the breath sounds are vesicular, and no ronchi and there is no wheezing, S1 single/S2 normal split regular, pansystolyc murmur was found, Extremities: warm acral, no clubbing finger, no edema extremities. The patient underwent a 12 lead ECG recording with Sinus Rhythm, regular, rate 70 bpm, right axis deviation, P mitral wave 0.010 sec, PR interval 0.16 sec, QRS duration 0.12 sec, R/S in V1 is greater than 1, with T inverted in lead V1-V3 (Figure 1).

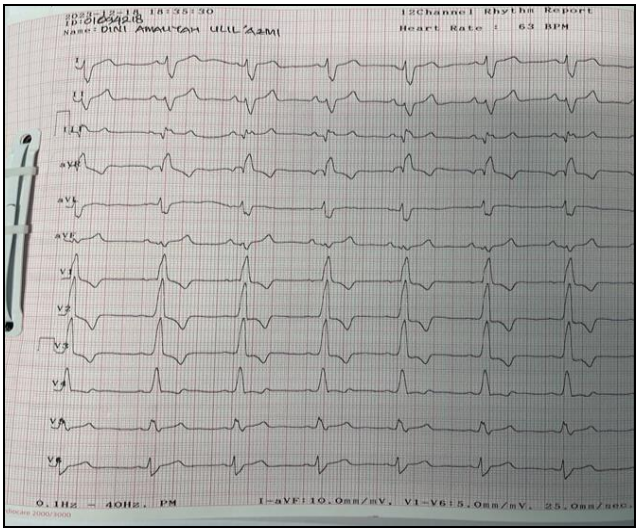

Fig 1: Electrocardiogram

Chest Xray examination we found cardiomegaly with intact sternal wire at CostovertebraeT4-T9. Laboratory investigation revealed within normal limits.

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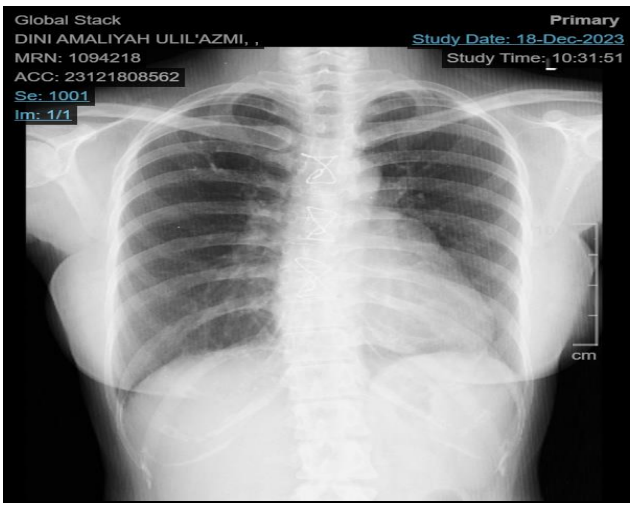

Fig 2: Thorax X-Ray

The echocardiogram examination revealed the presence of residual severe pulmonal stenosis following TOF repair, with a maximum pressure gradient (PS maxPG) ranging from 98-116 mmHg and a maximum velocity (PS Vmax) of 4.9 m/s. Additionally, there was severe pulmonal regurgitation, indicated by a pulmonary hypertension time (PR PHT) of 95 ms and a regurgitant jet width greater than two-thirds of the right ventricular outflow tract (RVOT). The examination also showed moderate to severe tricuspid regurgitation, with a tricuspid valve closure (TR VC) of 5.6 mm, a maximum velocity (TR Vmax) of 4.6 m/s, and a maximum pressure gradient (TR maxPG) of 85 mmHg. The right atrium and right ventricle were dilated, while the left ventricle had a D-shaped appearance and normal systolic function in both the left and right ventricles.

Fig 3: Echocardiography

On 2015 Patient was diagnosed ToF and underwent total repair surgery and found Cardiomegaly, normal LV contractility. Vena Innominate was present, diameter RPA 14 mm and LPA Ø 14 mm (HS: 14) confluence pulmonary artery. Ventricular septal defect perimembrane type size 20 mm, Overriding aorta, Main Pulmonary Artery diameter 16 (Full Size: 16.2). Pulmonary stenosis valvar dan subvalvar, Infundibular stenosis with single Ostium coronary transverse in RVOT.

In our hospital we then performed right heart catheterization and found Severe Pulmonary Stenosis with RV pressure 74/9(14), PA pressures 25/19 (9), RV-Pullback gradient 32/22 (14) mmHg , PA-Pullback gradient 25/19 (19) mmHg and then we calculate McGoon Ratio 1.52, Nakata Index 236, Half size 14, we found MAPCA at posterior AoD that connectong to right and left pulmo. After cardiac catheterization patient was in good condition and sent home and in queue for schedule for next surgical re-intervention.

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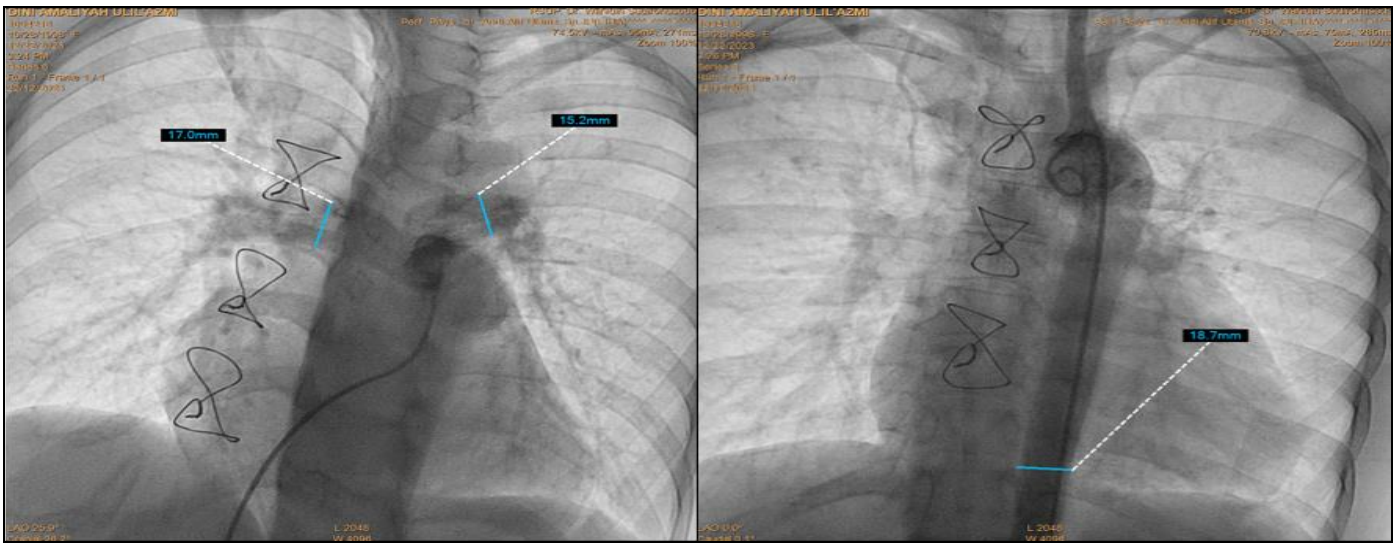

Fig 4: Right Heart Catheterization

IV. DISCUSSION

A. *Definition of Tetralogy of Fallot*

The prevalence of Tetralogy of Fallot is 5% to 10% of all cyanotic congenital heart defects and is the most common cyanotic heart disease. TOF occurs more in boys than girls and in all racial and ethnic groups. It is associated with several genetic conditions, including trisomy 21 (Down syndrome), and with deletions in chromosome 22 and can occur with other birth defects (myung). The ToF population can be divided into 2 groups. 1) ToF patients with syndromes account for around 20% (eg microdeletion 22q11, trisomy 21, Alagille, Noonan, William, and Klippel Fell) and 2) non-syndromic TOF patients (which represent the majority of TOF cases) ToF has 4 typical abnormalities consisting of heart and blood vessel defects (Park et al, 2020; Alboliras et al, 2018):

- Ventricular septal defect (VSD). There is a gap or defect in the ventricular wall;
- Pulmonary stenosis (PS). there is narrowing in the infundibular, valvar, supra-valvar or branches of the pulmonary artery (PA) or can be at multiple levels of narrowing;
- Overriding Aorta, Aortic Dextroposition (Ao);
- Right ventricular hypertrophy. The right ventricular wall is thicker than normal.

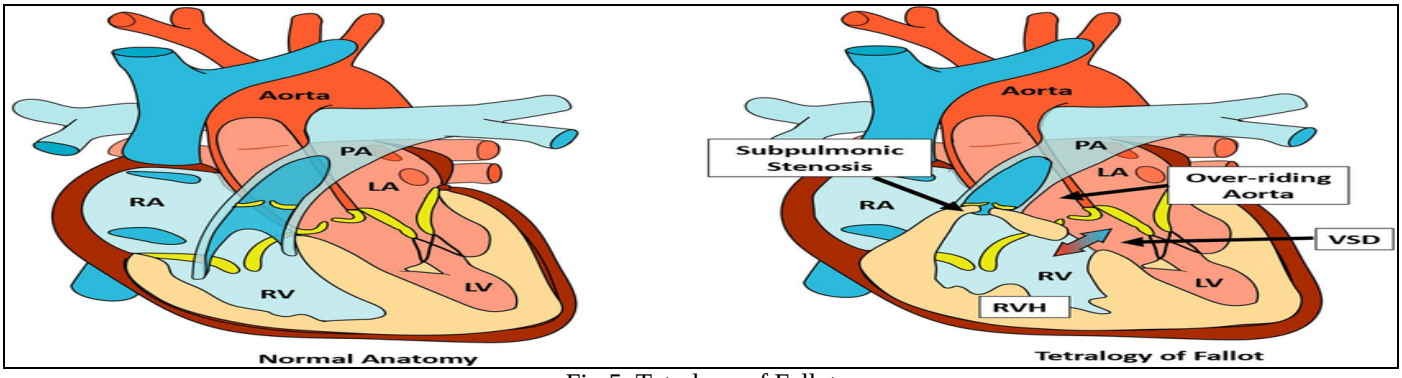

Fig 5: Tetralogy of Fallot

The VSD type in ToF refers to a significant defect located near the membrane with an expansion towards the subpulmonary region. The most common type of RVOT obstruction is infundibular stenosis, accounting for 45% of cases. Construction of the pulmonary valve is infrequent, occurring only in 10% of cases. There is also the possibility of a combination of both, which occurs 30% of the time. Pulmonary valve atresia is the most severe manifestation of an abnormality, occurring in 15% of cases. Most patients have a condition where the pulmonary annulus and major pulmonary artery are underdeveloped. The diameter of the branches of the pulmonary artery is typically small, although hypoplasia is infrequently observed. Pulmonary artery branch stenosis, particularly in the left pulmonary artery, is the most frequent occurrence. In certain instances, systemic collateral arteries may penetrate the lungs, particularly in cases of severe Tetralogy of Fallot (ToF) (Park et al, 2020; Alboliras et al, 2018; Mosa et al 2023). In 25% of cases, the aortic arch is situated on the right side.Based on the patient's history, physical examination, and supporting tests, it can be determined that the patient has a cyanotic kind of congenital cardiac disease known as Tetralogy of Fallot, which is the most common form of this condition.

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B. Morphology of Tetralogy of Fallot

The primary anomaly in TOF is the underdevelopment of the infundibular septum, which can result in the septum shifting forward and causing an anterior malalignment type ventricular septal defect (VSD). The crista supraventricularis is positioned in front of the parietal and septal bands, causing a constriction in the right ventricle outflow tract (RVOT). The medial papillary muscle is absent. A ventricular septal defect (VSD), typically of significant size, is located just behind the anteriorly displaced crista supraventricularis. The right sinus of Valsalva is situated at an elevated location in comparison to most hearts, allowing for easy access to the aorta from the right ventricle (RV). When there is significant or severe infundibular stenosis, the degree of overriding of the aorta is particularly pronounced. The pulmonary valve frequently exhibits stenosis. Frequently, it is bicuspid, and in 25% of cases, the pulmonary valve is atretic (Mosa et al, 2023; Vijayalakshmi et al, 2013).

Coronary anomalies are seen in 5 percent of cases, with the aberrant origin of the left anterior descending (LAD) artery from the right coronary artery (RCA) being the most frequently observed. An frequently observed anomaly is the anterior descending artery originating from the right coronary artery and running in front of the right ventricular outflow tract, which prevents surgical incision in this area (Alboliras et al, 2018).

As the aorta advances in relation to the pulmonary artery, the left main coronary artery must travel a greater distance in order to pass behind the pulmonary artery before dividing into the anterior descending and circumflex coronary arteries. As expected, a small percentage (approximately 5%) of patients have an abnormal distribution of their coronary arteries. In these cases, the anterior descending coronary artery originates from the right coronary artery and crosses over the infundibulum of the right ventricle before descending downwards in the anterior intraventricular groove. The circumflex coronary artery maintains its typical origin and courses posteriorly to the main pulmonary artery prior to entering the left atrioventricular groove. Infrequently, the right coronary artery originates from the left coronary artery and traverses towards the right over the infundibulum of the right ventricle. In the rare SDI variation of tetralogy, the right coronary artery is positioned in front of the infundibulum of the right ventricle. This variant is characterized by the primary pulmonary artery being located to the right of the aorta (Park et al, 2020; Mosa et al, 2023).

Upon surgical evaluation of our patient, we observed stenosis in the right ventricular outflow tract (RVOT), namely in the infundibular, valvar, and subvalvar regions. Additionally, we discovered a single coronary ostium in the RVOT, which was transverse in orientation and hindered transannular resection. This solitary ostium coronary corresponds to the typical patterns of coronary anatomy observed in Tetralogy of Fallot. Park et al stated that the existence of an anomalous coronary artery may cause a postponement of surgery until the child reaches one year of age, as it may need the use of a conduit between the right ventricle (RV) and the pulmonary artery (PA). Consequently, in our case, the infundibular resection for pulmonary stenosis cannot be performed optimally.

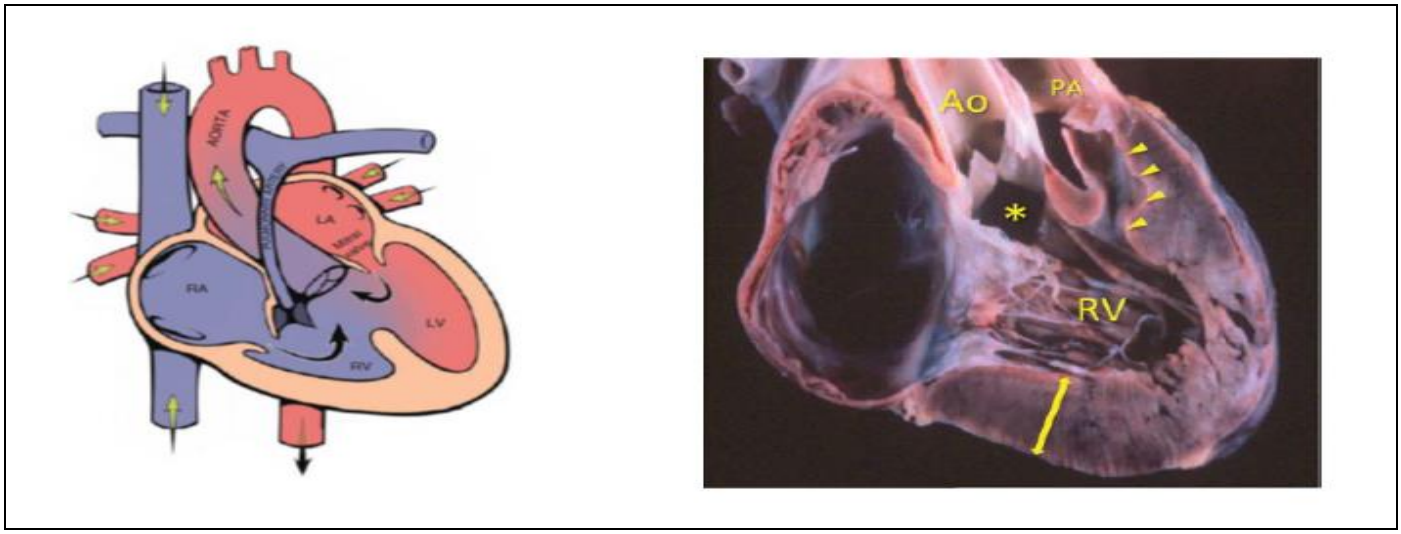

Fig 6: Anatomy Tetralogy of Fallot with Pulmonary Stenosis

As the aorta advances in relation to the pulmonary artery, the left main coronary artery must travel a greater distance in order to pass behind the pulmonary artery before dividing into the anterior descending and circumflex coronary arteries. It is not surprising that a small percentage (approximately 5%) of patients have abnormal coronary distribution, in which the anterior descending coronary artery originates from the right coronary artery and crosses the infundibulum of the right ventricle before descending downwards in the anterior intraventricular groove. The circumflex coronary artery maintains its typical origin and courses posteriorly to the main pulmonary artery before entering the left atrioventricular groove. Infrequently, the right coronary artery originates from the left coronary

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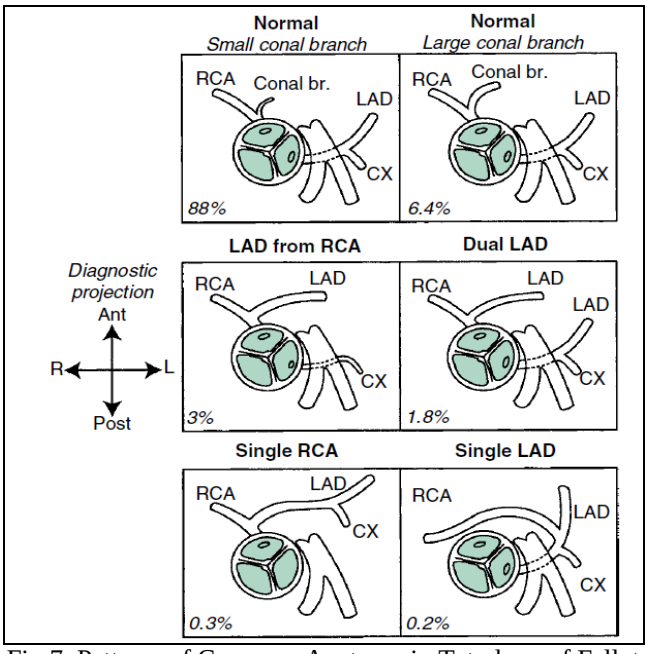

Fig 7: Patterns of Coronary Anatomy in Tetralogy of Fallot

In our patient, based on the results of surgery and when evaluating the RVOT, infundibular, valvar and subvalvar stenosis was found, which then after being released, a single coronary ostium was found in the RVOT which was transverse and obstructed transannular resection. This single ostium coronary match with patterns of coronary anatomy in Tetralogy of Fallot. Park et al said that The presence of anomalous coronary artery may delay the time of surgery until after 1 year of age because it may require a conduit between the RV and the PA, That's why in our patient the infundibular resection for pulmonary stenosis cannot optimal.

C. Morphology of Tetralogy of Fallot

The scope of medical intervention for Tetralogy of Fallot (TOF) is restricted. Cyanotic spells, also known as hypercyanotic spells, typically do not occur during the newborn period. The highest occurrence of these episodes is observed between the ages of 2 and 4 months. Oral beta-blockers are occasionally employed as a preventive measure against hypercyanotic episodes. Diuretic treatment and high calorie formula can effectively manage heart failure symptoms caused by pulmonary overcirculation in acyanotic TOF. Occasionally, oral propranolol medication is employed at a dosage of 0.5 to 2mg/kg every 6 hours to prevent hypoxic episodes. Although not commonly performed, balloon dilatation of the right ventricular outflow tract (RVOT) and pulmonary valve has been used as a technique to postpone the repair procedure for several months (Mosa et al, 2023; Vijayalakshmi et al, 2013).

Initial surgical therapy involves the creation of a systemic to pulmonary shunt in order to increase blood flow to the lungs. This is followed by a complete repair procedure. The shunt surgery typically involves the placement of a Gore-Tex tube that connects either the innominate artery (modified Blalock–Taussig shunt) or the ascending aorta (central shunt) to the pulmonary artery (Jonas, 2014).

The comprehensive repair entails the closure of the ventricular septal defect (VSD) using either a transatrial or transpulmonary approach, as well as the removal of muscle bundles in the right ventricular outflow tract (RVOT). In cases where the pulmonary valve and infundibulum are significantly underdeveloped, it may be essential to use a transannular patch to supplement them. As a result, the infant experiences a considerable amount of backflow of blood through the pulmonary valve, leading to the development of long-term issues such as enlargement and impaired functioning of the right ventricle. In the present period, there are attempts being made to conserve the original pulmonary valve, even if it means tolerating a moderate level of remaining pulmonary stenosis. This is done to prevent the long-term consequences of uncontrolled backflow. If an abnormal coronary artery is found in front of the right ventricular outflow tract (RVOT), it may be necessary to establish a connection between the right ventricle and the pulmonary artery (RV to PA conduit). This procedure is often done once the child reaches one year of age (Alboliras et al, 2018; Jonas, 2014).

Surgical palliative shunt procedures are performed to increasse pulmpnary blood flow, when the following situations are present, a shunt operation is usally performed rather than primary repair.

- Neonates Neonates with TOF and pulmonary atresia;
- Infants with hypoplastic pulmonary annulus, which requires a transannular patch for complete repair;
- Children with hypoplastic PAs;
- Unfavorable coronary artery anatomy;
- Infants younger than 3 to 4 months old who have medically unmanageable hypoxic spells;
- Infants weighing less than 2.5 kg.

There are several procedures were perfomed in the past, but Modified BT shunt is the procedure that performed at this time.

- Classic BT-Shunt: Anastomosed between the subclavian artery and the ipsilateral PA, is usually performed for infants older than 3 months because it is often thrombosed in young infants;
- The Potts Operation (1946): anastomosed between the descending aorta and the left PA, is no longer performed either.
- The Waterston Shunt (1962): anastomosed between the ascending aorta and the right PA, is no longer performed because of a high incidence of surgical complications resulting from this procedure included

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too large an shunt, leading to CHF or pulmonary hypertension, and narrowing and kinking of the right PA at the site of the anastomosis.

- Modified BT shunt: A Gore-Tex interposition shunt is placed between the subclavian artery and the ipsilateral PA. This is the most popular procedure for any age, especially for infants younger than 3 months of age (Park et al, 2020; Alboliras et al 2018; Jonas, 2014).

The defect is fully repaired by cardiopulmonary bypass, circulatory arrest, and hypothermia. The surgery involves closing the ventricular septal defect (VSD) by patching it, ideally using the transatrial and transpulmonary artery approach instead of the right ventriculotomy method. It also entails enlarging the right ventricular outflow tract (RVOT) by dividing or removing the infundibular tissue, and performing pulmonary valvotomy. The use of a transannular fabric patch is avoided. Expanding the right ventricular outflow tract (RVOT) without using a patch is more likely to be successful if the repair is performed during early infancy. Nevertheless, if the pulmonary annulus and major pulmonary artery are underdeveloped, the implantation of a transannular patch is inevitable. Certain medical institutes recommend the use of a monocusp valve during the initial repair procedure, while others suggest replacing the pulmonary valve at a later time if necessary (Vijayalakshmi et al, 2013; Jonas, 2014).

Table 1: Treatments Strategy for Tetralogy of Fallot with Pulmonary Stenosis

Table 1. Treatments for Tetralogy of Fallot With Pulmonary Stenosis

Age	Characteristics	Treatments
Newborn	Severe cyanosis in newborns at higher risk for complete repair: premature infants, infants with other birth defects, very ill infants	TEMPORARY PROCEDURES: SURGERY: Shunt procedure: Gore-Tex tube from an artery off the aorta to the pulmonary artery to provide blood flow to the lungs CATHETERIZATION: dilation of the pulmonary valve
3–6 mo	Open heart surgery, heart-lung bypass Type of surgical repair depends on child's anatomy	SURGERY: complete repair: 1. Close the VSD with a patch so that the aorta comes from the left ventricle 2. Remove extra-thick muscle in the right ventricle below the valve 3. Repair narrow pulmonary valve a. Valve-sparing technique for mild obstruction: pulmonary valve is dilated b. Transannular patch for severe obstruction: larger patch going across the pulmonary valve extending from the right ventricle to the pulmonary artery that destroys the pulmonary valve 4. Patch the right ventricle to relieve obstruction In 5% of patients, an abnormal coronary artery (supplies blood to heart muscle) crosses the area where the patch would go. In that case, repair is done with a conduit (tube) from right ventricle to main pulmonary artery
Adolescents and adults	Progressive leaking of the pulmonary valve and dilation of the right ventricle Narrow pulmonary valve or pulmonary artery	SURGERY: pulmonary valve replacement with tissue valve CATHETERIZATION: balloon dilate pulmonary valve or pulmonary artery

In our patient, the diagnosis of ToF was made when the patient was a teenager of 15 years, with repeated complaints of shortness of breath without signs of cyanosis, based on clinical status and supported by the results of an echocardiography examination that found a large VSD, overriding aorta >50%, pulmonary stenosis so it was decided to undergo total repair surgery.

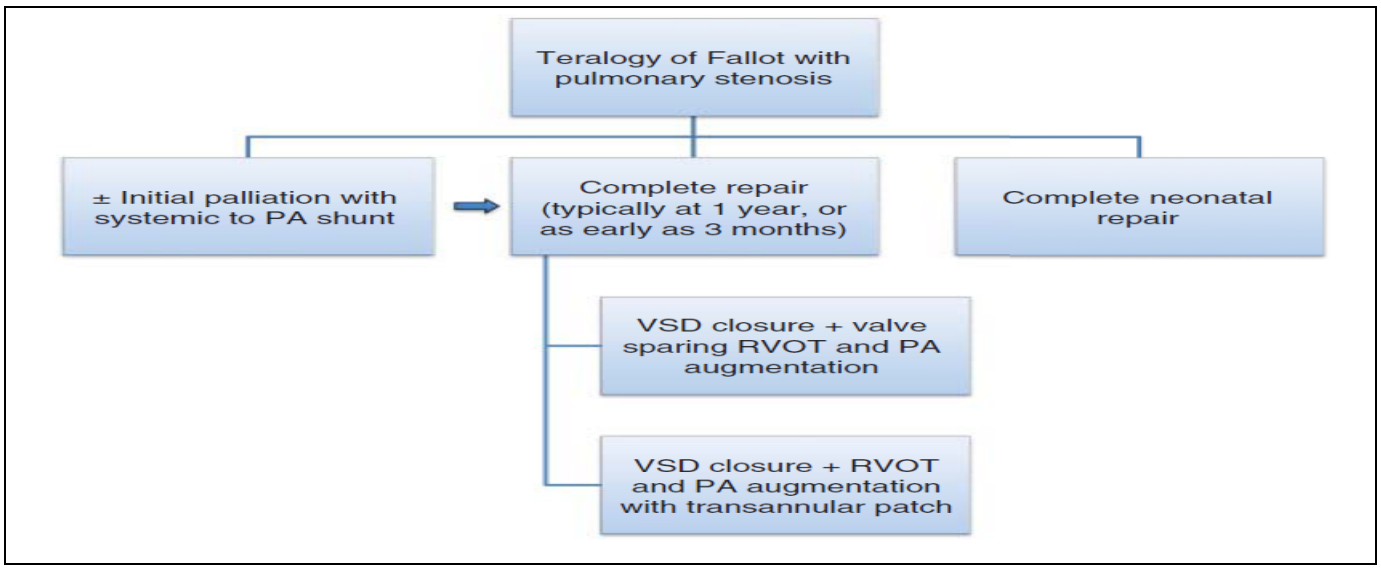

Fig 8: Surgical Management Options for Tetralogy of Fallot Patient

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D. Outcome

Potential long-term risks include the need for re-operation and valve replacement due to pulmonary valve regurgitation. Occasionally, immediate postoperative phase may present with total heart block. The survival rate in Tetralogy of Fallot (TOF) after surgical intervention exceeds 96% during the first two years of life and remains above 90% for a period of 30 years. Nevertheless, the long-term survival rate is lower compared to that of the general population. Atrial re-entrant tachycardia affects around 30% of patients, while significant ventricular arrhythmias occur in up to 10%. The annual occurrence rate of sudden death is 0.2% (Fujii et al, 2010; Sanchez et al, 2020).

Individuals who have had surgical repair for tetralogy of Fallot (TOF) have the capacity to live regular lives with exceptional heart function. The majority of survivors are classified as either New York Heart Association (NYHA) Class I or II. Certain people experience heightened symptoms with physical effort. Progressive pulmonary regurgitation (PR) occurs in 90 percent of individuals who have complete correction. Pulmonary regurgitation, which is often progressive, can occur following the correction of tetralogy of Fallot (TOF). While the patient may initially tolerate pulmonary regurgitation (PR) for a period of ten to twenty years, the presence of moderate to severe PR might eventually result in a substantial increase in volume inside the right ventricle (RV) and impaired RV function (Sanchez et al, 2020). It is advisable to undertake regular office examinations every 6 to 12 months for patients who have residual VSD shunt, residual obstruction of the RVOT, residual PA obstruction, arrhythmias, or conduction abnormalities, particularly for long-term monitoring. The proposed criteria for doing pulmonary valve replacement in patients who have previously undergone surgery for Tetralogy of Fallot (TOF) and have substantial pulmonary regurgitation (PR) are as follows. In order to use the proposal, it is necessary for the patient to have moderate to severe PR, with a regurgitation fraction of at least 25% in the right ventricle. The most effective method for studying RV function is through the use of MRI. However, if MRI is challenging or not recommended due to the presence of metallic objects or a cardiac pacemaker, a CT scan can be utilized as an alternative (Van Der Ven, 2019; Englert et al, 2019).

Table 2: Guidelines for ToF Recommendation

COR	LOE	Guidelines
I	B-NR	CMR can help quantify ventricular size and function, pulmonary valve function, pulmonary artery anatomy, and left-sided heart abnormalities in patients with repaired TOF.
I	B-NR	PVR (surgical or percutaneous) can relieve symptoms in patients with repaired TOF and moderate or greater PR with unexplained cardiovascular symptoms.
IIb	C-EO	PVR may be considered for preservation of ventricular size and function in asymptomatic patients with repaired TOF and ventricular enlargement or dysfunction with moderate or greater PR.
IIa	B-NR	Surgical PVR may be considered for adults with repaired TOF and moderate or greater PR, with other lesions requiring surgical interventions or ventricular tachyarrhythmias.

CMR = cardiac magnetic resonance; COR = Class of Recommendation; LOE = Level of Evidence; PR = pulmonary regurgitation; PVR = pulmonary valve replacement; TOF = tetralogy of Fallot.

In our patient, during the evaluation after surgery 8 years after, he still had complaints of intermittent chest pain, then echocardiography findings showed a severe pulmonary regurgitation with PHT 95 ms regurgitant jet width >2/3 RVOT and residual RVOT gradient 116 mmHg. Based on the Clinical Practice Guidelines for case management at the National Heart Center Hospital, post-TOF surgery patients with residual pulmonary stenosis need to undergo a right heart catheterization examination to assess the severity of the obstruction.

We performed right heart catheterization and found Severe Pulmonary Stenosis with RV pressure 74/9(14), PA pressures 25/19 (9), , RV-Pullback gradient 32/22 (14) mmHg , PA-Pullback gradient 25/19 (19) mmHg and then we calculated McGoon Ratio 1.52, Nakata Index 236, Half size 14, we found MAPCA at posterior AoD that connects to right and left pulmo. From echocardiography we found Residual Severe Pulmonal Stenosis post TOF Repair , (PS maxPG 98-116 mmHg, PS Vmax 4.9 m/s), Severe Pulmonal Regurgitation (PR PHT 95 ms, Regurgitant jet width >2/3 of RVOT) ,Moderate to Severe Tricuspid Regurgitation (TR VC 5.6 mm, TR Vmax 4.6 m/s, TR maxPG 85 mmHg) ,Right Atrium and Right Ventricle dilated with LV D-Shaped. Surgical intervention must be carried out if residual obstruction is found with RV systolic pressure > 2/3 of the systemic pressure in the form of residual infundibular, valvar or supravalvar PS, and said that it is necessary to carry out an MRI examination to measure RV LV volume and function, as well as the degree of PR for determine indications for pulmonary valve replacement.

E. Residual and Re-Interventions

Residual right ventricular outflow tract obstruction (RVOTO) frequently occurs after surgery and leads to the persistence or gradual increase of thickening of the right ventricle (RV). The findings from the INDICATOR study indicate that an enlarged right ventricle (RV) with a higher mass-to-volume ratio is a more significant long-term risk factor for ventricular tachycardia (VT) and mortality compared to the degree of RV dilatation (RV end-diastolic volume index). Existing recommendations offer explicit criteria for re-intervention in cases of residual right ventricular outflow tract obstruction (RVOTO). Valvular pulmonary stenosis (PS) can be treated with balloon valvuloplasty or percutaneous valvuloplasty (PVR) (Van Der Ven et al, 2019).

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Posterior resection (PR) is frequently observed during the medium- to long-term monitoring period. Between five to ten years following the repair, a significant proportion of patients, ranging from 40% to 85%, experience mild to severe PR. Pulmonary regurgitation (PR) causes an excessive amount of blood to flow into the right ventricle (RV), leading to the gradual enlargement of the RV. This enlargement may result in the occurrence of tricuspid regurgitation (TR) and impaired functioning of the RV. PVR is efficacious in reducing right ventricular (RV) volumes, mitigating tricuspid regurgitation (TR), diminishing QRS length, augmenting left ventricle (LV) ejection fraction (EF), and enhancing functional status. It is important to mention that there is currently no evidence to support the idea that PVR (Pulmonary Valve Replacement) improves survival compared to conventional therapy (Englert et al, Kim et al, 2013).

Table 3: Indications for Pulmonary Valve Replacement in Current Guidelines

	European Society of Cardiology (2010)[75]	American College of Cardiology/American Heart Association (2008)[73]	Canadian Cardiovascular Society (2009)[74]
Class I	Symptomatic patients with severe PR and/or PS (RV systolic pressure >60 mm Hg, TR velocity >3.5 m/s)	Severe PR and Symptoms or decreased exercise tolerance	
Class IIa	Severe PR or PS (or both) and either:	Severe PR and either:	Free PR and either:
RV size		Moderate to severe RV enlargement	EDVi 170 mL/m^2
Progression of RV size	Progressive RV dilation		Progressive RV dilation
RV function	Progressive RV dysfunction	Moderate to severe RV dysfunction	Moderate to severe RV dysfunction
TR	Progressive TR, at least moderate	Moderate to severe TR	Important TR
PS	PS RV systolic pressure greater than 80 mm Hg, TR velocity 4.3 m/s	Peak instantaneous echocardiography gradient greater than 50 mm Hg or RV/LV pressure ratio greater than 0.7 or Residual RVOT obstruction (valvular or subvalvular) with progressive and/or severe dilatation of the RV with dysfunction	RV pressure at least 2/3 systemic pressure
Exercise capacity	Decrease in objective exercise capacity		Symptoms such as deteriorating exercise performance
Arrhythmia	Sustained atrial or ventricular arrhythmia	Symptomatic or sustained atrial and/or ventricular arrhythmias	Atrial or ventricular arrhythmia

EDVi, end-diastolic volume index; LV, left ventricle; PR, pulmonary regurgitation; PS, pulmonary stenosis; RV, right ventricle; RVOT, right ventricle outflow tract; TR, tricuspid regurgitation.

V. CONCLUSION

This case report describes a 25-year-old lady who underwent surgical correction for Tetralogy of Fallot. Despite having the surgery 8 years ago, she was discovered to have residual severe pulmonary stenosis and severe pulmonary regurgitation during a follow-up examination. Alongside the discovery, there were reports of intermittent chest pain that was believed to be caused by damage to the coronary artery during a surgical procedure to remove a blockage in the pulmonary artery. Subsequent monitoring of elder groups has revealed the harmful consequences of PR over an extended period. Nevertheless, lingering lesions result in considerable morbidity. The transatrial and transpulmonary methods, as well as limited usage of TAPs, have been extensively utilized in surgical procedures to maintain pulmonary valve function. Although a significant number of adult ToF survivors will need one or more PVRs during their lives, it is still difficult to determine the best candidates and the best time for PVR. Gaining a deeper comprehension of RV failure becomes crucial in order to address these challenging inquiries. This could offer therapy alternatives to achieve best long-term health results for persons diagnosed with Tetralogy of Fallot (ToF).

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