Stent in the right ventricular outflow tract and delayed correction in a patient with tetralogy of Fallot: A clinical case.

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Abstract:

Introduction: Patients with symptomatic newborn tetralogy of Fallot indicate treatment, either palliative or corrective surgery; however, the initial management of these patients is currently controversial.

Clinical Case: Newborn female, 40 weeks gestation, who was admitted at 24 hours of life due to cyanosis, tachypnea, thoraco-abdominal retractions, and the presence of a III/VI systolic ejection murmur in the pulmonary focus.

Diagnostic workshop: In the tests: leukocytosis 27.05 x 103/UL, metabolic acidosis PH: 7.36, pCO2: 36.2 mmHg, Base excess: -4.6 mEq/L, hypoxemia: pO2 54.1 mmHg and lactic acidosis five mmol/L. The echocardiogram showed tetralogy of Fallot, with severe infundibular and pulmonary valve stenosis, with a 3-mm (Z: -7.6) pulmonary ring, hypoplastic pulmonary trunk, and branches.

Treatment: Prostaglandin, ampicillin, and IV gentamicin were started for seven days. On the 11th day, cardiac catheterization was performed with stent placement (4.5 and 16 mm) in the right ventricular outflow tract, with five high-flow aortopulmonary collaterals embolized.

Evolution: The procedure was well tolerated, with discharge after 17 days with acetylsalicylic acid 15 mg/day, furosemide 2 mg/day, and spironolactone 6.25 mg/day. At ten months, corrective surgery for tetralogy of Fallot was performed with a transannular patch and plasty of the left pulmonary branch. The control at 18 months was standard.

Conclusion: Placing a stent in the outflow tract of the right ventricle percutaneously is a safe method that guarantees stable pulmonary flow, allowing corrective surgery to be deferred outside the neonatal stage.

Keywords: **MeSH**: Tetralogy of Fallot, Newborns, Cyanosis, Stents, Case Reports.

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Introduction

Tetralogy of Fallot is the most frequent cyanotic congenital cardiopathy, representing between 5 and 10% of congenital cardiopathy [1, 2].

Neonates with tetralogy of Fallot, who are severely symptomatic, are indicated to undergo immediate treatment, whether soothing or corrective. Managing these patients generates controversy since repair in the neonatal stage is not possible in all centers. Different studies have shown higher mortality, with a longer hospitalization time and a higher incidence of reinterventions [3- 6].

Three months is now considered a good age for repair of patients with Tetralogy of Fallot [5], although many groups have reported good results with earlier ages. Performing a systemic pulmonary fistula is the usual management of these patients. Nevertheless, the procedure is not exempt from morbidity, mortality, and complications with the possible risk of deformation of the pulmonary branches [3]. Blalock-Taussig shunt mortality ranges in neonates from 9 to 11% [7, 8].

Placing a stent in the right ventricular outflow tract is a suitable option that guarantees stable pulmonary flow, favors the development of pulmonary branches, and allows elective corrective surgery to be performed without the difficulties that reoperation offers [5, 6, 9].

Clinical case

Clinical history

This is a female newborn, 40 weeks gestation, with an adequate weight for gestational age of 3.2 kg and a height of 52 cm, with a prenatal history of premature rupture of membranes obtained by cesarean section. The patient was admitted 24 hours after extrauterine life and was referred from another care center due to cyanosis and tachypnea crises.

Labial and nail cyanosis (acrocyanosis) and thoracoabdominal retractions were observed on physical examination. On auscultation, the presence of a rude III/VI systolic ejection murmur in the pulmonary focus was striking, with an audible pulmonary murmur without added noises; capillary refill was 3 seconds, with a respiratory rate of 65 per minute, oxygen saturation 80% with oxygen by nasal cannula, and heart rate of 138 beats per minute.

Diagnostic workshop

Laboratory tests showed leukocytosis 27.05 x 10^3/UL, with a predominance of neutrophils 17.45 X 10^3/UL, with normal PCR and elevated procalcitonin 2.28 ng/ml, compensated metabolic acidosis PH: 7.36, pCO_2: 36.2 mmHg, Base excess: -4.6 mEq/L, low oxemia: pO_2 54.1 mmHg and high lactic acid of 5 mmol/L. The rest of the laboratory tests were normal.

Chest X-ray showed preserved lung volumes with a boot-shaped cardiac silhouette.

Abdominal and transfontanellar ultrasounds were reported as usual.

The transthoracic echocardiogram showed tetralogy of Fallot, with severe infundibular and pulmonary valve stenosis, a pulmonary annulus of 3 mm (Z: -7.6), a hypoplastic pulmonary trunk and branches (3 mm each branch), a gradient of 50 mmHg, and a subaortic ventricular septal defect wide, aortic overhang, right ventricular hypertrophy, patent foramen ovale, restrictive atypical ductus arteriosus versus aortopulmonary collateral, adequate ventricular function, ejection fraction 57%, shortening fraction 29%, usual emergency coronary arteries.

Treatment

Initial management is carried out with intravenous hydration and initiation of prostaglandin, reversing acidosis, and improving pressure and saturation. In infectious cases, it is managed with ampicillin for ten days and gentamicin for seven days due to a history of premature membrane rupture.

On the 11th day of hospitalization and with standardized tests, therapeutic cardiac catheterization was performed with stent placement (4.5 and 16 mm) in the right ventricular outflow tract plus embolization of 5

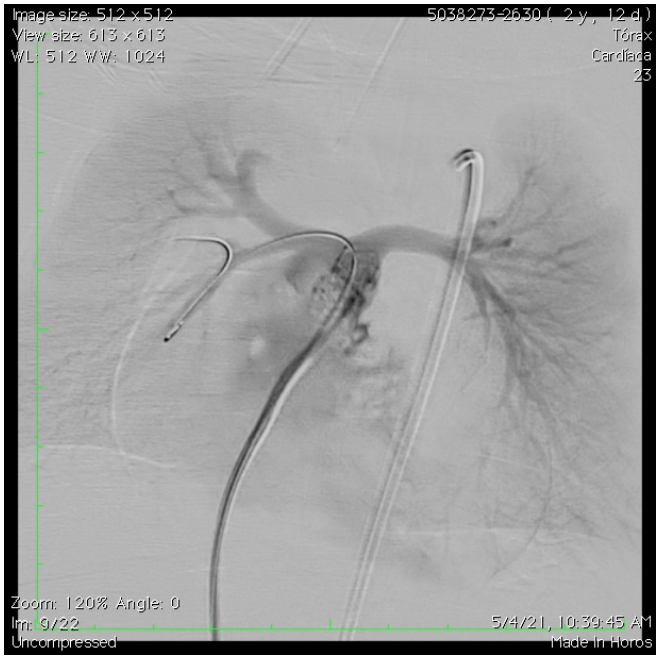
high-flow aortopulmonary collaterals. The stent was placed via the femoral route and covered the ventricular outflow tract, the annulus, and part of the pulmonary trunk (see Figure 1). It was insufflated at 8 atmospheres with an estimated 5.2 mm.

The procedure was well tolerated, and the patient was extubated the next day with oxygen saturation above 90%. The control echocardiogram showed a stent in the patent right ventricular outflow tract, continuous pulmonary branches, hypoplastic right 3.3 mm (Z: -2), and aortopulmonary collaterals.

Evolution

He was discharged 17 days after hospitalization with acetylsalicylic acid 15 mg/day, furosemide 2 mg/day, spironolactone 6.25 mg/day, and outpatient follow-up by cardiology.

Figure 1. Cardiac catheterization with right ventricular outflow tract stenting.



Cardiac catheterization with anteroposterior projection stented right ventricular outflow tract. The flow of permeable contrast medium into the pulmonary branches is observed. Pulmonary branches of suitable diameter without stenosis.

A follow-up echocardiogram at four months of age showed a stent treating the right ventricular outlet and pulmonary ring with a maximum gradient of 90 mmHg, confluent pulmonary branches, good development of 5 mm each, and aortopulmonary collaterals.

At ten months of age, a new diagnostic and therapeutic catheterization was performed, which reported severe tetralogy of Fallot with stented right ventricular outflow tract, pulmonary trunk and branches with sound development, slight stenosis of the origin of the left pulmonary branch, and six high-flow aortopulmonary collaterals embolized with microcoil and solid devices.

Finally, corrective surgery for Tetralogy of Fallot was performed six days after the last catheterization. Tetralogy of Fallot correction was performed with a transannular patch plus left pulmonary branch plasty. The procedure was well tolerated, without incident, and the patient was discharged on the 7th day of surgery.

Postoperative echocardiogram reported closed VSD, mild pulmonary stenosis, a maximum gradient of 20 mmHg, pulmonary branches without stenosis, good ventricular function, and an ejection fraction of 67% (Figure 2). Echo monitoring of the pulmonary branches is described in Table 1.

Table 1. Growth of the pulmonary branches in follow-up by echocardiography.

Age	24 hours	2 weeks	4 months	18 months
Weight (kg)	3.2	3.2	6.02	10.88
Size (cm)	52	fifty	61.5	81
RPB in mm (Z)	3 (Z= -2.70)	3.3 (Z= -2.04)	5 (Z=-0.94)	9 (Z=-0.8)
LPB in mm (Z)	3 (Z= -2.23)	3.6 (Z= -1)	5 (Z= -0.34)	8 (Z= -0.84)

RPD; Right pulmonary branch, RPI; Left pulmonary branch, (Z) Z value indexed by weight and height, whose zero value is normal.

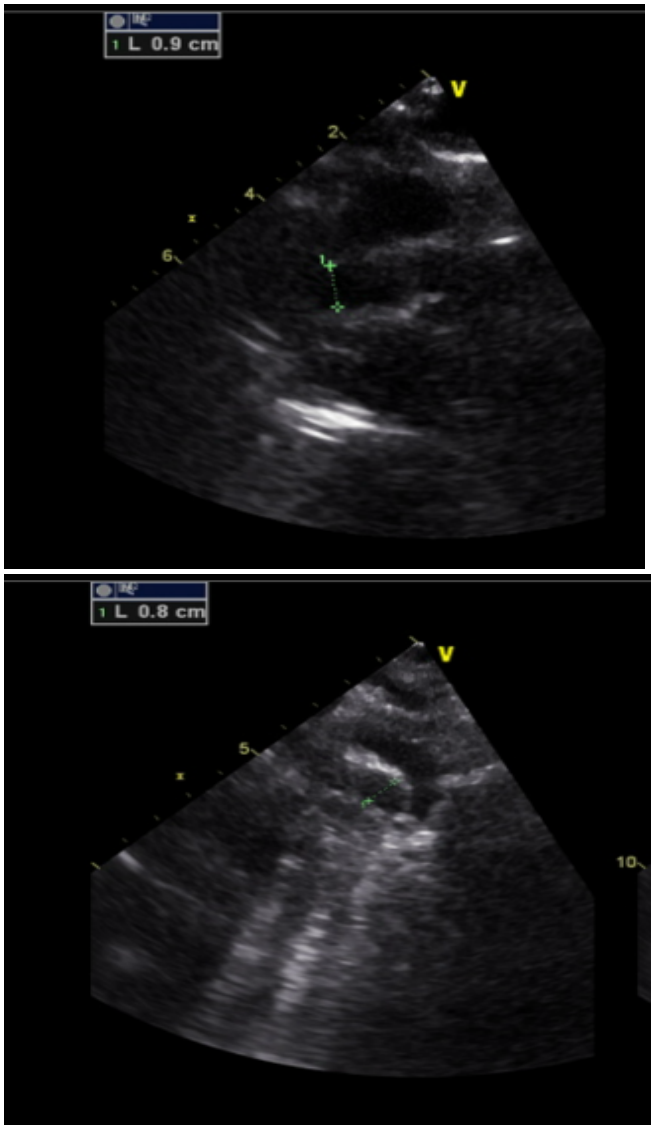
Discussion

Newborn patients with symptomatic tetralogy of Fallot with poor anatomy represent a real challenge for the medical team. Using prostaglandin to keep the ductus patent is the initial step in managing these patients until

the medical-surgical unit proposes the best therapeutic strategy according to the possibility of the center where the patient is located.

The frequently used strategy is the creation of a systemic pulmonary fistula, either with the creation of a Blalock-Taussig shunt or a central aortopulmonary shunt. However, these procedures are not exempt from mortality, which is between 9 and 11%; they are generally difficult to manage postoperatively [3, 5, 7, 8]. Added to all this is the risk of a reintervention during delayed correction.

Figure 2. Ultrasound of pulmonary arteries.



Suprasternal axis: 9 mm right pulmonary branch is observed; left pulmonary branch 8 mm.

Some series are described with good results in primary correction of tetralogy of Fallot at increasingly earlier ages; however, in the neonatal stage, the outlook still needs to be clarified since there is an increase in mortality, reoperations, and longer length of stay of hospitalization in these patients, [3- 6, 10] even more so, if it is associated with hypoplasia of pulmonary branches as in the case presented.

The latter was demonstrated in a meta-analysis conducted by Loomba in 2017 [11], where the results obtained with surgical correction in the neonatal stage versus repair outside the neonatal stage were compared.

Stenting in the ductus arteriosus and the right ventricular outflow tract constitutes an excellent alternative for these patients. Both procedures guarantee a stable pulmonary flow and allow the development of the pulmonary branches [5, 6, 9, 12]; however, they may not apply to all patients. Atypical ducts or ducts with complex anatomy are not easily dilatable with stents, and a malpositioned stent can obstruct or deform a pulmonary branch. On the other hand, the stent in the right ventricular outflow tract is only indicated if the pulmonary valve is hypoplastic and, therefore, cannot be preserved in corrective surgery, as is the case in the present patient.

The outflow tract stent proved to be a safe method in several studies, reducing the need for intensive therapy and hospitalization time compared to the Blalock Taussig shunt in a survey by Quandt in 2017.

Conclusions

In neonates with severe tetralogy of Fallot, symptomatic dependence on ductus arteriosus, and severely hypoplastic pulmonary valves, stent placement in the right ventricular outflow tract is a safe method. It constitutes an excellent alternative to initial palliative treatment and to defer corrective surgery outside the neonatal period without the complications of a reoperation.

Abbreviations

VSD: Intraventricular communication.

Supplementary information

No supplementary materials are declared.

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Does not apply.

Author contributions

Darwin Zhune Villagrán: Conceptualization, Data conservation, Acquisition of funds, Research, Resources, Software, Writing - original draft.
Simón Duque Solorzano: Conceptualization, Data conservation, Supervision, Acquisition of funds, Research, Resources.
Boris Barreno Martínez: Conceptualization, Supervision, Acquisition of funds, Investigation, Resources.
Paola Mendieta Chispe: Research, Software, Writing - original draft.
All the authors have read and approved the final version of the manuscript.

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Surgical procedures, laboratory studies, and imaging were part of the regular activities of the pediatric surgery service and were not an additional cost to the patient. The researchers covered the administrative costs of this investigation.

Availability of data and materials

The data sets generated and analyzed during the current study are not publicly available due to clinical case confidentiality.

Statements

Ethics committee approval and consent to participate

Not required for clinical cases.

Publication Consent

The authors have permission for publication from the patient's parents.

Conflicts of interest

The authors declare they have no conflicts of interest.

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