ORIGINAL ARTICLE

Immediate and Mid-term Outcomes of Blalock-Taussig Shunt versus Right Ventricular Outflow Tract Stenting in Symptomatic Neonates with Tetralogy of Fallot

Comparación de resultados inmediatos y a mediano plazo de la anastomosis de Blalock-Taussig versus la colocación de stent en tracto de salida de ventrículo derecho en neonatos sintomáticos con tetralogía de Fallot

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ABSTRACT

Background: The initial management of symptomatic neonate patients with tetralogy of Fallot (TOF) associated with unfavorable anatomy and significantly reduced pulmonary vascular flow is controversial and continues to be a clinical challenge.
Objective: The aim of this study was to describe the clinical evolution and to compare branch pulmonary artery growth in symptomatic neonatal TOF patients who received a modified Blalock Taussig (mBT) shunt versus right ventricular outflow tract stent placement (RVOTs) at the Department of Cardiology, Hospital de Niños de Córdoba, between March 2011 and March 2021.
Methods: A retrospective, observational study identified 113 patients with TOF, 20 of which (18%) were symptomatic neonates requiring initial palliative intervention. Categorical variables are expressed as percentage and continuous variables as median and interquartile range (IQR). A p value <0.05 was considered significant.
Results: Among the 20 patients included in the study, 11 (55%) formed the mBT shunt group and 9 (45%) the RVOTs group. In the mBT shunt group, pre-palliation right pulmonary artery (RPA) Z score was -3 (IQR 4.20) and increased to -1.6 (IQR 1.56) (p= 0.11), and left pulmonary artery (LPA) Z score of -2.5 (IQR 4.8) increased to -1.80 (IQR 2.36) (p= 0.44). In the RVOTs group, RPA Z score prior to the palliative procedure was -3.45 (IQR 3.83) and increased to -2.5 (IQR 3.58) (p= 0.021) and LPA Z score of -4.10 (IQR 2.51) increased to -2.00 (IQR 3.75) (p= 0.011). Pre-intervention O2 saturation of 75% (IQR 6) increased to 87% (IQR 9) in the mBT shunt group (p= 0.005) and from 75% (IQR 16) to 91% (IQR 13) in the RVOTs group (p= 0.008). Mean length of hospital stay after the procedure was 10 days (IQR 11) in the mBT shunt group and 6 days (IQR 2) in the RVOTs group (p= 0.095).
Conclusions: In symptomatic neonates with TOF, both palliative strategies improved the clinical condition. In patients who received RVOTs, there was greater branch pulmonary artery growth. A larger number of cases and longer-term follow-up will be necessary to confirm these findings.

Key words: Tetralogy of Fallot - Stent - Right ventricular outflow tract - Pulmonary branches - Blalock-Taussig shunt

RESUMEN

Introducción: El manejo inicial de neonatos con tetralogía de Fallot (TF), con síntomas asociados a una anatomía desfavorable y un flujo vascular pulmonar significativamente reducido es controversial, y un desafío clínico.
Objetivo: Describir la evolución clínica y comparar el crecimiento de ramas pulmonares en neonatos con TF sintomáticos ingresados al Departamento de Cardiología del Hospital de Niños de Córdoba, desde marzo de 2011 hasta marzo de 2021, que recibieron anastomosis de Blalock-Taussig modificada (aBTm) versus colocación de stent en tracto de salida de ventrículo derecho (sTSVD).
Material y métodos: Estudio retrospectivo, observacional. Se identificaron 113 pacientes con TF; 20 de ellos (18%) fueron neonatos sintomáticos y requirieron paliación inicial. Las variables categóricas se expresan como porcentaje; las continuas como mediana y rango intercuartilo (RIC). Un valor de p <0,05 se consideró significativo.
Resultados: De los 20 pacientes incluidos en el estudio, 11 (55%) constituyen el grupo aBTm y 9 (45%) el grupo sTSVD. En el grupo aBTm la rama pulmonar derecha (RPD) pre paliación tenía un score Z -3 (RIC 4,20), que aumentó a -1,6 (RIC 1,56) (p= 0,11) post intervención; y la rama pulmonar izquierda (RPI) un *score* Z -2,5 (RIC 4,8) que se incrementó a -1,80 (RIC 2,36) (p= 0,44). En el grupo sTSVD la RPD pre paliación tuvo un score Z -3,45 (RIC 3,83) que aumentó a - 2,5 (RIC 3,58) (p= 0,021) y la RPI un score Z -4,10 (RIC 2,51) que se incrementó a -2,00 (RIC 3,75) (p= 0,011). La saturación de O2 (SO2) pre intervención fue 75% (RIC 6), y aumentó a 87% (RIC 9) en el grupo aBTm (p= 0,005); e inicialmente fue 75% (RIC 16) y aumentó a 91% (RIC 13) en el grupo sTSVD (p= 0,008). La mediana de estadía hospitalaria post procedimiento fue 10 días (RIC 11) en el grupo aBTm, y 6 (RIC 2) en el grupo sTSVD (p= 0,095).

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Conclusiones: En neonatos con TF sintomáticos, ambas estrategias paliativas mejoran la condición clínica. En los que recibieron sTSVD, se objetivó un crecimiento mayor de las ramas pulmonares. Mayor número de casos y seguimiento más largo serán necesarios para confirmar estos hallazgos.

Palabras claves: Tetralogía de Fallot - Stent - Tracto de salida de ventrículo derecho - Ramas pulmonares - Anastomosis de Blalock-Taussig

INTRODUCTION

The initial management of symptomatic neonates with Tetralogy of Fallot (TOF), associated with unfavorable anatomy and significantly reduced pulmonary vascular flow, is controversial and is still a clinical challenge.

Neonatal primary surgical repair is infrequent due to its early high morbidity and mortality and often need of reoperations. Alternatively, modified Blalock-Taussig (mBT) systemic-pulmonary shunt has been used for decades to increase pulmonary blood flow, reduce hypoxemia and promote branch pulmonary artery growth. This strategy allows definite surgical repair to be performed at a more advanced age, generally between 3 and 11 months of age, according to the clinical evolution of the patient. (1)

Frequently, mBT shunt as initial strategy in these early symptomatic patients is associated with elevated indices of complications, including acute obstructions, distortion of branch pulmonary arteries, hyperfunction and occasionally mortality. (2) Moreover, few data have been reported in the literature evaluating branch pulmonary artery growth after mBT shunt.

Conversely, stent implantation to relieve the right ventricular outflow tract (RVOT) obstruction avoids a palliative and/or corrective surgery in the neonatal period of a critical patient, has the potential to stimulate better branch pulmonary artery growth and prevents their distortion compared with a mBT shunt intervention. However, the native pulmonary valve is often covered by the stent, and thus with this technique, a transannular patch would be necessary after stent removal during the complete surgical repair. (3-7)

The aim of the present study was to analyze the clinical evolution and branch pulmonary artery growth of symptomatic neonates with TOF receiving mBT shunt versus RVOT stenting (RVOTs).

METHODS

An observational, retrospective study was carried out in symptomatic neonate and infant patients under one year of age, with diagnosis of TOF, admitted to the Department of Cardiology of Hospital de Niños de la Santísima Trinidad de Córdoba from March 2011 to March 2021. All patients received palliative treatment as initial approach to their heart disease. The decision regarding initial palliation was agreed by the cardiovascular team during clinical-surgical seminars. On admission, all patients presented unfavorable anatomy (branch pulmonary arteries with Z score ≤ -2.5), cyanotic crises (oxygen saturation <70%), cardiogenic shock, low weight, prematurity or other significant comorbidities (necrotizing enterocolitis, kidney failure, and/or convulsions) that decided an initial percutaneous or surgical palliative approach.

The Z score for each cardiac structure reported was obtained according to the age, weight and height of each patient. (8) Echocardiographic measurements of the branches of the pulmonary arteries were performed immediately before the initial palliative procedure, and were repeated one month before the final corrective surgery to assess their growth. Oxygen saturation by pulse oximetry (sO_2) and length of hospital stay after the procedure were recorded. Finally, age, palliation time and surgical technique used (transannular patch / preservation of the pulmonary annulus) at the time of complete surgical repair were also registered.

Statistical analysis

Categorical variables are expressed as percentage and continuous variables as median and interquartile range (IQR). The chi-square test or Fisher's test was used to compare categorical variables, the Wilcoxon signed-rank test to compare paired samples and the Mann-Whitney test to compare independent continuous samples. A p value <0.05 was considered significant. SPSS24 software package was used for the statistical analysis.

Ethical considerations

The palliative approach used, either mBT shunt or RVOTs was approved by the institutional Ethics Committee and the parents or legal guardians responsible for the patients signed the specific informed consent form to carry out the procedure.

RESULTS

Among a total of 113 patients identified with TOF, 20 (18%) were symptomatic neonates requiring initial palliation. Eleven patients (55%) received mBT shunt and the remaining 9 (45%) RVOTs. All presented cyanotic crises and 8 were receiving prostaglandins. Median age of the mBT shunt group at the time of the intervention was 60 days (IQR 75) and median weight 4.5 kg (IQR 3), while in the RVOTs group median age was 19.5 days (IQR 15) and median weight 2.95 kg (IQR 1.65). Three patients of the mBT shunt group presented complications: two with acute shunt occlusion and one developed hemopericardium with signs of cardiac tamponade, while in the RVOTs group no adverse events were recorded (p=0.28). No significant changes were found in branch pulmonary artery dimensions in the mBT shunt group: in the right pulmonary artery (RPA), the pre-palliation Z score was -3 (IQR 4.20), and increased to-1.6 (IQR 1.56) after the intervention (p=0.11), and in the left pulmonary artery (LPA), the pre-palliation Z score was -2.5 (IQR 4.8) and increased to -1.80 (IQR 2.36) after the procedure (p=0.44). In the RVOTs group, a significant difference was found in these dimensions: in the RPA, the pre-palliation Z score was -3.45 (IQR

3.83) and improved to -2,5 (IQR 3.58) after the intervention (p=0.021), and in the LPA the pre-palliation Z score was-4.10 (IQR 2.51) and increased to -2.00 (IQR 3.75) after the procedure (p=0.011) (Figures 1 and 2). A significant improvement was observed in sO_2 in both groups: median sO2 pre-intervention was 75% (IQR 6) and increased to 87% (IQR 9) after the procedure in the mBT shunt group (p=0.005) and from 75% (IQR 16) to 91% (IQR 13) in the RVOTs group (p=0.008) (Figure 3). Mean length of hospital stay after the procedure was 10 days (IQR 11) in the mBT shunt group and 6 days (IQR 2) in RVOTs group (p=0.095). Mean age at corrective surgery was 24 months (IQR 12) for the mBT shunt group and 7 months (IQR 4) for the RVOTs group. Definite surgical repair in the mBT shunt group required placement of a conduit between the right ventricle and the pulmonary artery due to coronary anomaly in 18% of patients, the pulmonary annulus was preserved in 27% and transannular patch placement was required in 55% of cases. In the RVOTs group, stent removal was possible in all patients, and the 9 cases that received this initial palliative technique required transannular patch placement. No deaths were recorded both in the palliative and corrective procedures in all the cohort of patients.

DISCUSSION

Most asymptomatic patients with TOF that present good-sized confluent branch pulmonary arteries, receive primary surgical repair between 3 and 9 months of age and weighing approximately 5-9 kg, with excellent results. (1, 2) Newborns or younger infants requiring earlier interventions, usually depend on prostaglandins or present severe cyanosis with very reduced pulmonary antegrade flow, as a consequence of unfavorable RVOT (infundibular or valvular stenosis) and/or pulmonary vascular tree anatomy (hypoplastic branch pulmonary arteries or presence of collateral vessels). An initial mBT shunt procedure is an effective strategy in these symptomatic patients and/or with poor anatomy or significant comorbidities (prematurity, low weight, infections, and neurological/renal/gastrointestinal abnormalities), even though it is associated with high early or late morbidity. (3) Remote complications, as branch pulmonary artery distortion, vocal cord or hemidiaphragm paralysis, hyperfunction, and secondary complications to thoracotomy/sternotomy, have a long-term impact. (4)

Percutaneous interventional techniques in congenital cardiac diseases have had an exponential development during the last 20 years. As a consequence of this progress, cyanotic premature and neonatal patients have been treated with stent implantation in the ductus arteriosus or in a dysfunctional mBT shunt to maintain adequate effective pulmonary vascular flow. Based on these experiences, RVOTs as alternative to mBT shunt is being chosen as a safe, effective and reproducible initial strategy in a growing number of patients. (6, 7, 9-11) Mid-term RVOTs results

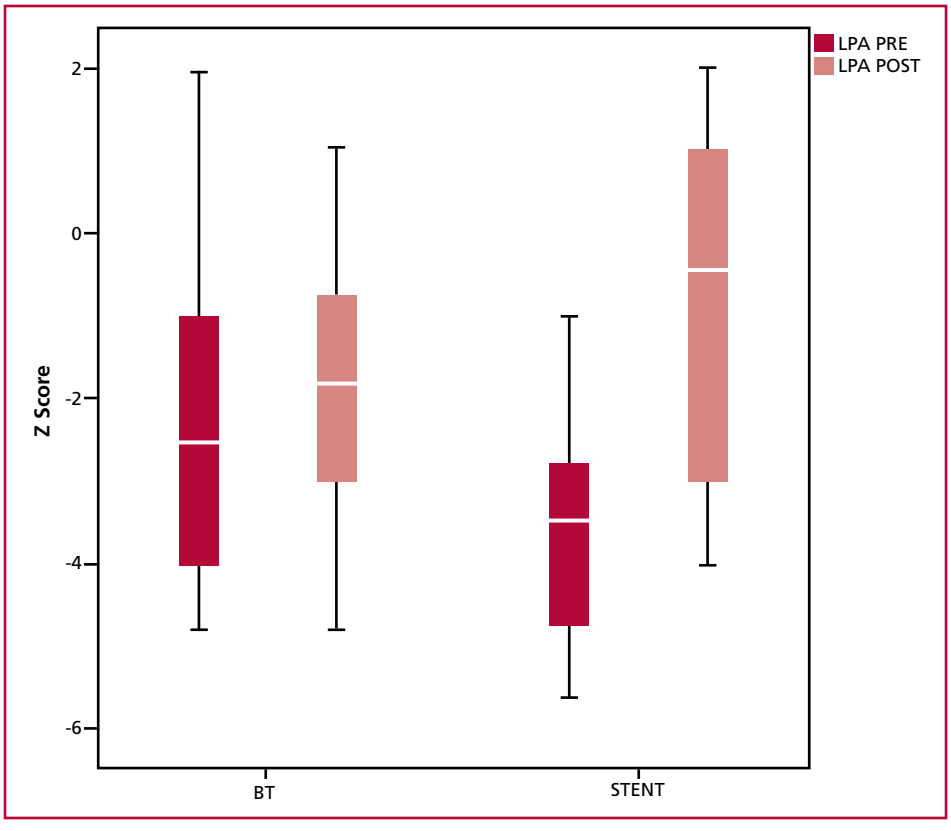

Fig. 1. Pre-and post-intervention Z score of the left pulmonary artery according to the selected palliative strategy: Blalock-Taussig shunt vs. stent placement in the right ventricular outflow tract. LPA: Left pulmonary artery BT: Blalock.Taussig.

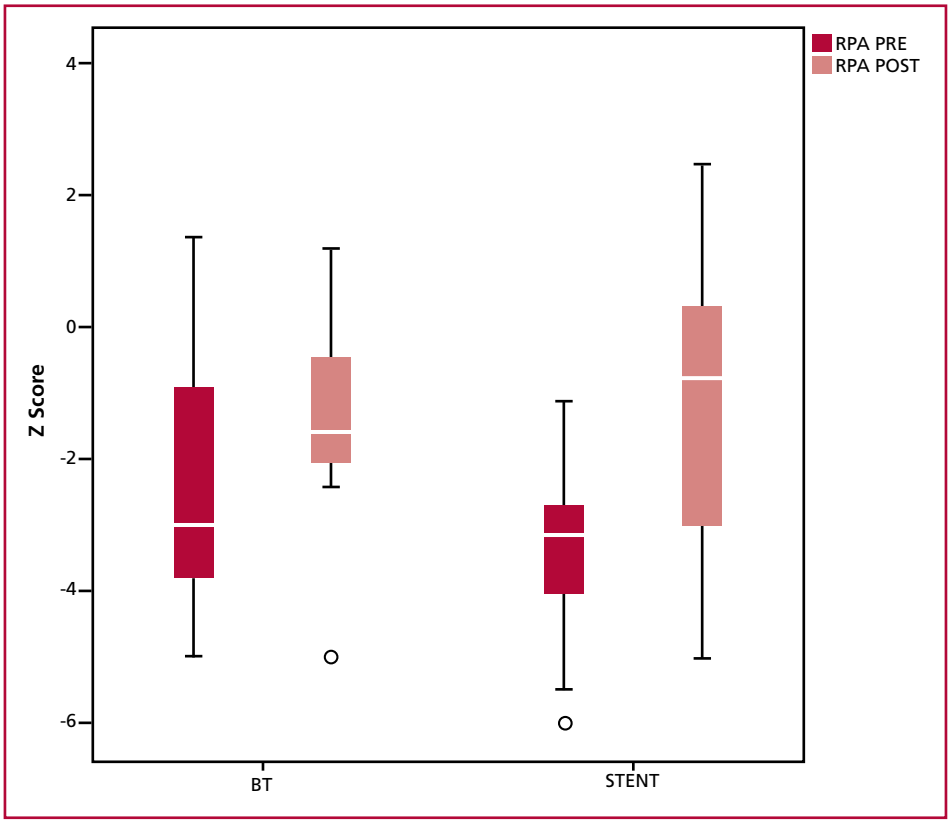

Fig. 2. Pre-and post-intervention Z score of the right pulmonary artery according to the selected palliative strategy: Blalock-Taussig shunt vs. stent placement in the right ventricular outflow tract. RPA: Right pulmonary artery BT: Blalock.Taussig

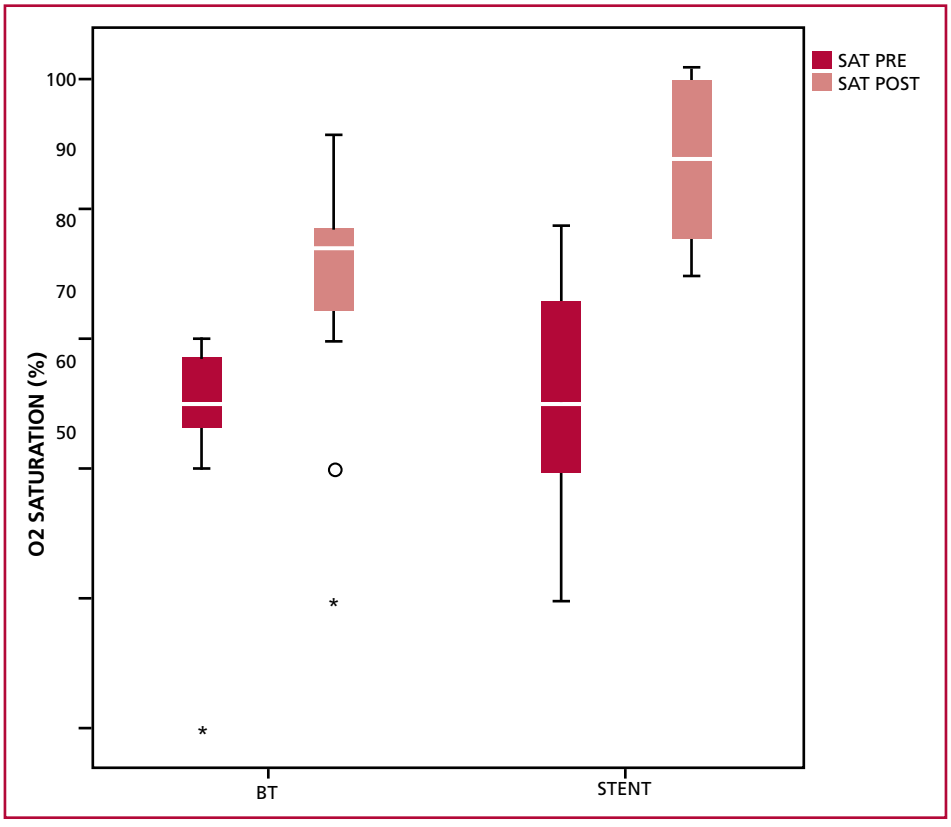

Fig. 3. Pre and post palliative intervention peripheral oxygen saturation assessed by pulse oxymetry in both groups. STENT: Stent in the right ventricular outflow tract, BT: Blalock-Taussig, SAT: Peripheral oxygen saturation

have shown similar survival to mBT shunt, with lower length of hospital stay and peri-procedural complications, though associated with a higher rate of reintervention than the surgical procedure. (5)

Our initial experience included seriously ill patients who presented with severe cyanosis, hypoxemia, were dependent on mechanical respiratory assistance, were receiving prostaglandins, and had very small branch pulmonary arteries as shown by their Z score. These patients were considered at "high risk" for any surgical procedure. Our hypothesis was that with this percutaneous technique a better and more stable hemodynamic condition is achieved, as the systemic venous return is directed to the pulmonary circulation, resulting in increased peripheral oxygen saturation. In addition, it prevents a reduced aortic diastolic perfusion pressure, leading to improved coronary perfusion, as opposed to what is observed with the mBT shunt procedure.

Surgical thoracotomy/sternotomy is also avoided, often requiring cardiopulmonary bypass with the potential complications it entails, especially in the neonatal period. (6) According to published reports, also branch pulmonary artery growth (Nakata and McGoon angiograplic indices and echocardiographic measurements) is greater with increased antegrade flow to the pulmonary vascular bed achieved by RVOTs, as opposed to flow provided by a mBT shunt (7, 12, 13)

The selection of initial palliative strategy using RVOTs over mBT shunt afforded a general benefit in our cohort, in terms of branch pulmonary artery growth with respect to that measured before corrective surgery, reduced acute complications and lower length of hospital stay. Time to complete definite surgical repair was remarkably shorter in the RVOTs group compared with the mBT shunt group. We believe that this is not only related to greater branch pulmonary artery growth, but also to a closer follow-up of patients who underwent this new palliative technique in our institution, and the trend to perform a complete repair at an earlier age to facilitate stent removal. There was a significant difference in the use of transannular patch during definite surgical repair, since in all patients with RVOTs, the stent went through the pulmonary valve and hence, patch placement was the rule. Lower length of hospital stay is explained by the less invasive percutaneous procedure compared with the initial palliative surgical approach, which in our institution is performed with sternotomy and frequently, with cardiopulmonary bypass.

Potential limitations can influence our results. Firstly, it was a retrospective, single-center study, which may imply biases at the time of treatment selection for each patient. Right ventricular outflow tract stenting was the initial palliation strategy chosen during the last three years.

No inter or intra-observer variability study was implemented for branch pulmonary artery measurement; however, this was performed by three expert operators with more than 10-year experience in the specialty. Both length of hospital stay as complication occurrence tended to be more favorable in the RVOTs group, although they did not reach statistical significance, possibly due to lack of statistical power. The present study reproduces experiences reported by other investigators on the efficacy and safety of RVOTs as bridge to corrective surgery. Prospective and/or randomized studies, with a larger number of patients will be necessary to identify the characteristics of patients who might benefit from one or the other palliation strategy in neonates with TOF evidencing early symptoms.

CONCLUSION

In this case cohort study, symptomatic neonates with TOF receiving RVOTs as initial palliation showed greater branch pulmonary artery growth compared with the mBT shunt group. Use of transannular patch at the time of definite surgical correction was imperative in the RVOTs group. Longer follow-up and a larger number of patients will be necessary to demonstrate clinical benefits.

Conflicts of interest

None declared.

(See authors' conflict of interests forms on the web/Additional material.)

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