EDITORIAL

Right Ventricular Outflow Tract Stenting as an Option to Blalock-Taussig Shunt for Tetralogy of Fallot

Implante de stent en el tracto de salida del ventrículo derecho como alternativa a la anastomosis de Blalock-Taussig en la tetralogía de Fallot

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Tetralogy of Fallot (TOF) is a complex cardiac condition that includes a wide anatomical spectrum. Surgical repair may have an uneventful outcome between 6 months and the first year of life in most patients with favorable anatomy (e.g. adequate size of branch pulmonary arteries). However, management remains a challenge in symptomatic patients that present early in life, often due to a more severe involvement of right sided obstruction. In this setting, cyanosis becomes evident as a consequence of the progressive narrowing of the pulmonary infundibulum as well as an increased right ventricular stroke volume that faces a semi-fixed resistance and directs flow towards the systemic circulation. In addition, these patients often show underdevelopment of right-sided structures, including a smaller pulmonary annulus, a short and tapered main pulmonary artery, as well as variable degree of diffuse hypoplasia and/or stenosis of the pulmonary arteries. (1) Thus, it is not surprising that most of these children exhibit early progressive cyanosis and are at potential risk of "hypoxic spells" that require early medical stabilization with prostaglandin infusion to maintain arterial duct patency until a more definitive therapy can be established.

Today, although some selected centers advocate for early surgical repair within the first 3 months of life (provided anatomical features are considered favorable), many countries (including Latin America) continue to support an initial palliative approach. Creation of a systemic-to-pulmonary shunt, usually a modified Blalock- Taussig type (MBTS) followed by complete repair at a later stage is still the most adopted strategy in many centers around the world. Far from being considered an ideal palliative measure, a MBTS can result in disproportionate growth of the pulmonary arteries or stenosis of the involved pulmonary branch that may hinder results at the time of complete repair. (2, 3) Furthermore, according to a recent publication, shunt thrombosis or pulmonary over-circulation can reach up to 23% and 30% respectively, along with up to a 12% in-hospital mortality in patients with ductal dependent pulmonary circulation undergoing a MBTS. (4)

The latter has long been considered a matter of genuine concern and has motivated clinical cardiologists, interventionists and even cardiovascular surgeons (as is the case in our center) to seek for non-surgical alternatives to MBTS in order to provide temporary relief of cyanosis and delay surgery. In the context of TOF with pulmonary atresia, ductal stenting may be considered a valuable option provided the ductal anatomy is favorable. (5) Alternatively, pulmonary valve balloon dilation may be considered in patients with TOF, if obstruction (usually mixed) predominates at the valvar level. This option, however, is currently seldom used due to the transient and non-sustained benefit in arterial oxygen saturation. In recent years, right ventricular outflow tract stenting has been established as an attractive palliative alternative to early repair and MBTS. (6, 7)

The case series presented in this issue of the Argentine Journal of Cardiology by Dr. Peirone et al. (8), whom we acknowledge and extend our congratulations, adds to the valuable list of publications that position right ventricular outflow tract stenting as a serious contender of the ideal palliative strategy in the context of early symptomatic TOF infants to maintain adequate pulmonary flow and allow surgical repair at a later stage in more favorable conditions. The authors share their experience consisting of six children (including three within the neonatal period) with history of severe cyanosis or hypoxic spells that underwent right ventricular outflow tract stenting as an alternative to MBTS. We believe some relevant observations are worth pointing out. Half of the patients in this series weighed less than 3 kg, including a nine-day-old weighing 2,200 g, which illustrates the possibility to perform this intervention safely and effectively even

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in a delicate subgroup of patients (i.e. prematurity, low weight or non-cardiac associated comorbidities). Also, the authors nicely demonstrate how both the pulmonary annulus and the branch pulmonary arteries were severely hypoplastic, possessing Z-score values well below a threshold that would be considered at all feasible to perform complete repair, but even low enough to perhaps convince a cardiovascular surgeon of performing a MBTS in the setting of a poor vascular pulmonary tree anatomy. The latter, is in our opinion, one of the greatest strengths of right ventricular outflow tract stenting over a MBTS. Stent placement in the pulmonary infundibulum allows continuous antegrade flow towards the pulmonary circulation, stimulating symmetrical and balanced growth of both pulmonary arteries. Quandt et al. (9) demonstrated infundibular stenting promoted greater increments in arterial saturations and pulmonary artery growth compared to patients undergoing MBTS. Furthermore, the Toronto group (10) nicely demonstrated right ventricular outflow tract stenting not only promoted significant pulmonary growth, but it also allowed patients that were initially considered of high-surgical risk to reach complete repair a few months later with overall good clinical outcomes comparable to those with an initial favorable cardiac anatomy that underwent repair within the first 3 months of life.

Finally, a word of caution is warranted for clinicians and interventionists considering right ventricular outflow tract stenting. Fully covering the entire length of the infundibulum is desirable to maintain adequate oxygen arterial saturation and prevent cyanosis. A drop in oxygen saturation might also occur in the presence of instent stenosis that might raise the need to either redilate the originally implanted stent or place additional stents to address this issue. Similarly, there is still debate about whether the stent should be extended beyond the pulmonary valve and into the main pulmonary artery or be deployed just below the valvar plane to potentially preserve the valve at the time of surgery. In this regard, we agree with the authors' decision to extend the stent across the pulmonary valve. The possibility to spare the pulmonary valve, is in our opinion, a complicated if not impossible undertaking if one is to consider that that these patients have often extremely hypoplastic pulmonary annulus. These patients will almost invariably require a transannular patch repair that can be extended into the main pulmonary artery and even single or bilateral branch pulmonary artery plasty in a good number of cases. (10)

As previously mentioned, we believe the authors have contributed to a valuable list of publications that have positioned right ventricular outflow tract stenting as a valid option for the early management of symptomatic TOF infants. We anticipate that far from being abandoned, right ventricular outflow tract stenting will be adopted by an increasing number of cardiovascular teams around the world who take care after children born with TOF and their variants with the goal to improve survival and quality of life, which fortunately enough, most of these patients already enjoy.

Conflicts of interest

None declared.

(See authors' conflicts of interest forms on the website/Supplementary material).

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