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Fate of Hypoplastic Pulmonary Arteries After Arterial Duct Stenting in Congenital Heart Disease With Duct-Dependent Pulmonary Circulation

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ABSTRACT

OBJECTIVES This study sought to evaluate the impact of arterial duct (AD) stenting in promoting catch-up growth of hypoplastic pulmonary artery (PA) tree in congenital heart disease with duct-dependent pulmonary circulation (CHD-DPC).

BACKGROUND Significant and balanced PA growth following AD stenting has already been consistently reported in the literature. However, no data are so far available about the role of this approach in severe PA hypoplasia, which significantly impacts the risk of surgical repair.

METHODS Pre-surgical angiographic PA evaluation was performed in 45 patients with confluent PAs submitted to neonatal AD stenting as palliation of CHD-DPC. PA growth was evaluated as Nakata Index and McGoon ratio as well as individual PA *z*-score changes, both in the whole population and according to the original vessel size (Nakata Index <100 mm^2/m^2, Group I [n $=$ 15] vs. Nakata Index >100 mm^2/m^2, Group II [n $=$ 30]).

RESULTS Control angiography was performed 7.5 $\pm$ 6.5 months (median 6 months) after duct stenting, showing significant and balanced PA growth. The Nakata Index increased from 143 $\pm$ 73 mm^2/m^2 to 270 $\pm$ 88 mm^2/m^2 (124 $\pm$ 118%, $p < 0.0001$); left PA *z*-score from -0.7 ± 1.7 to 1.0 ± 1.4; right PA *z*-score from -0.6 ± 1.3 to 1.2 ± 1.3 ($p < 0.0001$ for both comparisons). Group I showed a greater increase of global PA growth (Nakata Index increase 227 $\pm$ 141% vs. 72 $\pm$ 57%, $p < 0.001$) as compared with Group II. Final PA size did not significantly differ between the groups (246 $\pm$ 105 mm^2/m^2 vs. 282 $\pm$ 78 mm^2/m^2, p $=$ NS).

CONCLUSIONS Percutaneous AD stenting is highly effective in promoting a significant and balanced catch-up growth of diminutive PAs, being therefore advisable in this subset of patients as a reliable alternative to surgical palliation.
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Arterial duct (AD) stenting is widely considered an effective palliation of congenital heart disease with duct-dependent pulmonary circulation (CHD-DPC) in high-risk patients unsuitable for primary repair (1-6). This option may promote significant and more balanced pulmonary artery (PA) growth as compared with surgical systemic-to-pulmonary artery shunt (7-10). However, no study has so far specifically addressed the fate of hypoplastic PAs following AD stenting, which may significantly impact the overall risk of surgical repair.

The aim of this study was to evaluate the role of AD stenting in promoting the growth potential of diminutive PAs in patients with CHD-DPC.

METHODS

PATIENT POPULATION. Between April 2003 and December 2014, 118 neonates and infants with

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CHD-DPC underwent successful AD stenting at our institution as a cost-effective alternative to surgical palliation. Mean hospital stay for AD stenting was 12 days in a regular ward compared with 10 days (3 days in a post-surgical intensive care unit and 7 in a regular ward) for a surgical shunt, resulting in a similar global economic impact (€9,823 vs. €11,090). Among these patients, 45 needed just a short time of support to pulmonary circulation, 17 are still in follow-up before planned surgical repair, and 4 died before hospital discharge. The remaining 52 patients needed surgical repair over a mid-term follow-up and were submitted to control cardiac catheterization 7.5 ± 6.5 months (range 2 to 45 months, median 6 months) after the initial palliation. Seven patients were excluded from the analysis because the stented duct supplied disconnected PAs. The demographic and clinical data of the remaining 45 patients are reported in Table 1. At the time of AD stenting, 39 patients showed biventricular physiology, and the remaining 6 patients were supposed to be candidates for Fontan track. Thirteen patients had complete duct dependency of the pulmonary circulation due to trivial or absent additional pulmonary blood flow (PBF) (28.9%), whereas the remaining 32 patients showed mild, but clinically insignificant, accessory PBF. Eight of the 12 patients with pulmonary valve atresia with intact ventricular septum and none of the patients with pulmonary valve atresia with ventricular septal defect had been submitted to pulmonary valve perforation at the time of AD stenting. No patient showed hemodynamic significant aortopulmonary collaterals at control angiography. At the time of AD stenting, 15 patients showed very hypoplastic PAs (Nakata Index <100 mm^2/m^2, range 46 to 100, median 79 mm^2/m^2; Group I), whereas the remaining 30 patients had normal or mildly reductive PAs (Nakata Index >100 mm^2/m^2, range 110 to 399, median 164 mm^2/m^2; Group II). No difference was found between groups in terms of intracardiac anatomy, as well as complete duct dependency of the pulmonary circulation (Table 1).

INTERVENTIONAL PROCEDURE. AD stenting was performed under general anesthesia following a previously described protocol, with the aim of covering the entire AD length (4–6). Mean prostaglandin dosage at the time of AD stenting was 0.02 g/kg/min, but prostaglandin infusion had been stopped some few hours before the procedure. The stent size was individually chosen on the basis of patient size, ductal anatomy, and expected time for which palliation was needed. However, it was usually about 25% smaller than the planned surgical shunt in the belief that it acted more as a central shunt than a Blalock-Taussig shunt. After stent deployment, repeat aortic angiograms were performed in multiple views to exclude incomplete coverage of the duct as well as to evaluate the PA size and any potential stent-related PA stenosis. After the procedure, long-term acetylsalicylic acid treatment was planned at a dose of 3 to 5 mg/kg/day.

ABBREVIATIONS AND ACRONYMS

AD = arterial duct

CHD-DPC = congenital heart disease with duct-dependent pulmonary circulation

PA = pulmonary artery

PBF = pulmonary blood flow

ANGIOGRAPHIC MEASUREMENTS. Control cardiac catheterization was scheduled whenever oxygen saturation was consistently reduced over at least 2 follow-up visits or before scheduled surgical repair. Pulmonary angiography was performed in right anterior oblique and four-chamber views (Figure 1), measuring individual PAs at the site of their first branching point. The diameter of the descending

TABLE 1 Clinical, Demographic, and Angiographic Data of Patients Submitted to Control Angiography Following AD Stenting

N = 45 (complete duct-dependent pulmonary circulation 28.9%)	
Age, months	8.7 ± 7.2 (range 3–45)
Weight, kg	6.2 ± 2.6 (range 2.5–15)
Diagnosis	
Critical PS/PA-IVS	12
ToF	12
PA-VSD	9
TV Ebstein's anomaly	1
cTGA with critical PS/atresia	2
TA	1
TGA-VSD with critical PS/atresia	6
UVH with critical PS/atresia	1
Criss-cross heart with ToF	1
Group I (Nakata Index <100 mm^2/m^2, n = 15)	
Complete duct-dependency: 37.5%	
Control angiography, months: 10 ± 11 (median 6.5)	
Critical PS/PA-IVS:	3
ToF:	5
PA-VSD:	4
TV Ebstein's anomaly:	1
cTGA with critical PS/atresia:	2
Group II (Nakata Index >100 mm^2/m^2, n = 30)	
Complete duct-dependency: 24.1%	
Control angiography, months: 8 ± 5 (median 6)	
Critical PS/PA-IVS:	9
ToF:	7
PA-VSD:	5
TA:	1
TGA-VSD with critical PS/atresia:	6
UVH with critical PS/atresia:	1
Criss-cross heart with ToF:	1

Values are n unless otherwise indicated.

AD = arterial duct; cTGA = corrected transposition of great arteries; PA-IVS = pulmonary valve atresia with intact ventricular septum; PA-VSD = pulmonary valve atresia with ventricular septal defect; PS = pulmonary valve stenosis; TA = tricuspid valve atresia; TGA = transposition of great arteries; ToF = tetralogy of Fallot; TV = tricuspid valve; UVH = univentricular heart; VSD = ventricular septal defect.