

Transcatheter Stenting of Arterial Duct in Duct-Dependent Congenital Heart Disease

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SUMMARY

Introduction Critical congenital heart diseases (CHD) are mostly duct-dependent and require stable systemic-pulmonary communication. In order to maintain patency of the ductus arteriosus (DA), the first line treatment is Prostaglandin E1 and the second step is the surgical creation of aortic-pulmonary shunt. To reduce surgical risk in neonates with the critical CHD, transcatheter stenting of DA can be performed in selected cases.

Case Outline A four-month old infant was diagnosed with the pulmonary artery atresia with ventricular septal defect (PAA/VSD). The left pulmonary artery was perfused from DA, and the right lung through three major aortopulmonary collaterals (MAPCAs). A coronary stent was placed in the long and critically stenotic DA, with final arterial duct diameter of 3.5 mm, and significantly increased blood supply to the left lung. After the procedure, the infant's status was improved with regard to arterial oxygen saturation, feeding and weight gain. During the follow-up, one year later, aortography revealed in-stent stenosis. The left pulmonary artery, as well as the branches, was well-developed and the decision was made to proceed with further surgical correction.

Conclusion Stenting of DA can be an effective alternative to primary surgical correction in selected patients with duct-dependent CHD.

Keywords: congenital heart disease; duct dependent; transcatheter interventional procedure; stenting arterial duct

INTRODUCTION

Early definitive repair and less-invasive procedures are current trends in the management of congenital heart defects (CHD). Conventional shunt surgery in duct-dependent CHD in neonates and during early infancy is associated with significant morbidity and complications, such as shunt stenosis/thrombosis, pulmonary overflow with the pleural effusions, diaphragm paralysis, and distortion of pulmonary artery (PA) branches. Progress in the development of endovascular stent and implantation technique has enabled efficient alternatives to conventional surgical aortopulmonary shunts [1]. Stenting the persistent ductus arteriosus (DA) ensures its permanent patency and good pulmonary blood flow with favorable clinical response.

CASE REPORT

The complex cyanotic CHD was recognized in the first week of life when diagnosed, by echocardiography, with the pulmonary artery atresia with ventricular septal defect (PAA/VSD), an open ductus arteriosus with diameter of about 2 mm in connection with hypoplastic left pulmonary artery, and numerous major aortopulmonary collateral arteries (MAPCAs) in the right lung. The presence of large MAPCAs in the right lung enabled relatively satis-

factory arterial oxygen saturation (SatO₂) of 85% and stable general condition, without the need for the introduction of prostaglandin E1. Genetic tests have confirmed 22q11.2 micro-deletion.

During the follow up, at the age of three months, significant lowering of arterial SatO₂, to 55%, with a failure to thrive was identified. Heart catheterization corroborated formerly established diagnosis of PAA/VSD with DA diameter of approximately 0.5 mm at its narrowest point. Aortography clearly revealed that the left PA was supplied from DA (Figure 1), the right lung through three MAPCAs (Figure 2), and the right aortic arch with the mirror-image branching of vessels. On the basis of cardiac catheterization findings, a Cardiosurgical Board decided that an aortopulmonary shunt operation would involve a high level of risk, due to the hypoplastic left pulmonary artery and the left subclavian artery (diameter: 3 mm). Arterial duct stenting was suggested as an alternative palliative treatment.

Stenting technique: A 4-month-old female infant, weighing 3.5 kg, was re-catheterized using the trans-femoral approach, under general anesthesia. Antibiotic prophylaxis and Heparin 50 U/kg were administered at the beginning of the procedure. Aortography showed a small, 16 mm long DA extending at a very sharp angle from the left brachiocephalic artery: the narrowest point of DA was approximately 0.5 mm

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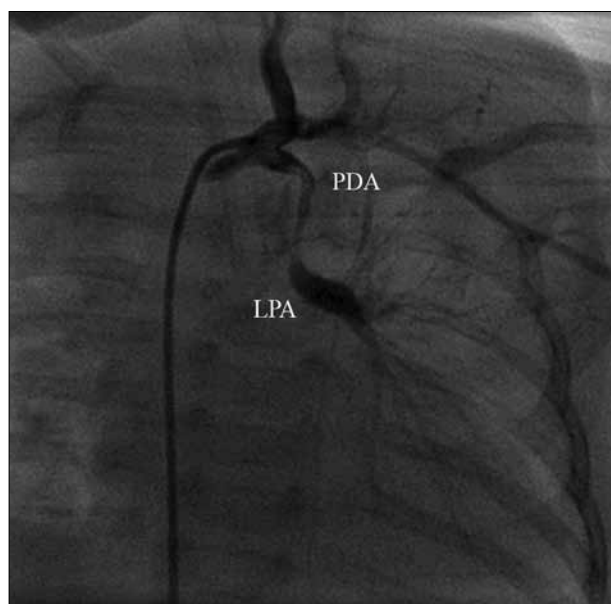


Figure 1. Arteriography: long and critically stenotic (arrow) patent arterial duct (PDA) from the left brachiocephalic trunk connected with the hypoplastic left pulmonary artery (LPA)

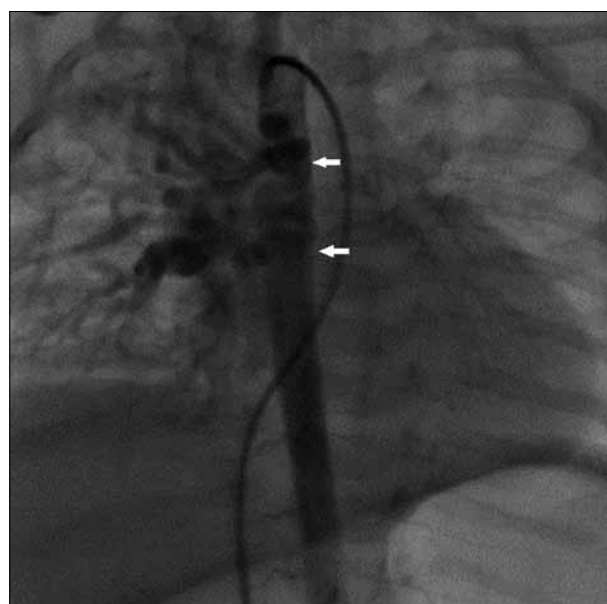


Figure 2. Descending aortography: right aortic arch with multiple aortopulmonary collaterals for the right hemithorax

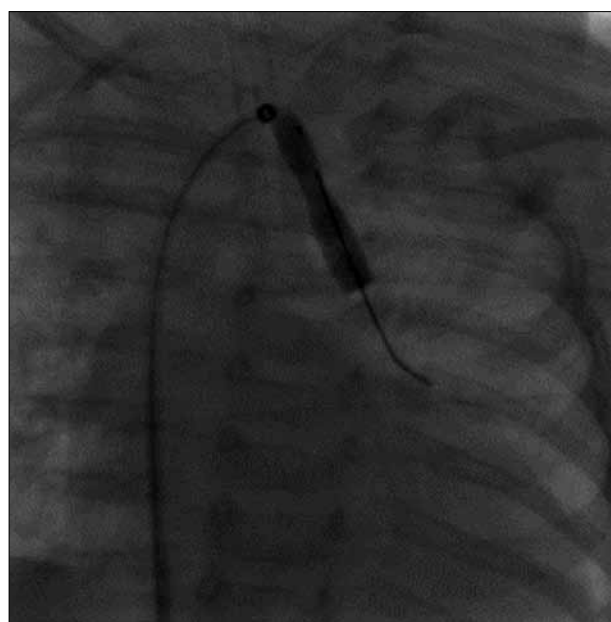


Figure 3. Stent implantation in the arterial duct with mild narrowing of the middle part

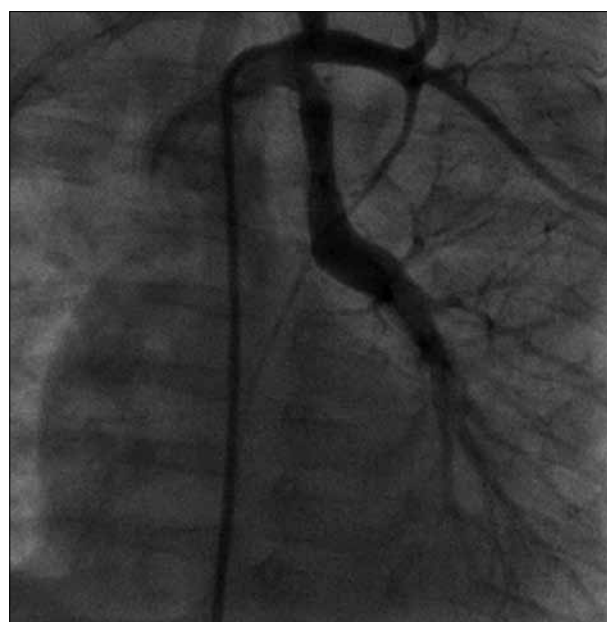


Figure 4. Significantly increased flow through ductal stent in the left pulmonary artery

at the pulmonary artery end. The left PA was poorly-developed at just 3 mm in diameter. After placement of the 0.014-inch coronary wire deep in the left PA, a 5 Fr guiding catheter was introduced through the DA to the left PA and, finally, the coronary stent was introduced. An 18 mm long, 4.5 mm diameter stent was chosen in order to cover the whole length of the DA (Cordis, BX VELOCITY 4.50×18 mm). After inflating the balloon catheter, we recorded a final stent diameter of 3.5 mm, which had a mild waist at the junction of DA and the left PA (Figure 3). Removal of deflated balloon from the stent was difficult, as a result of the sharp angle of the guiding catheter, but was successful without stent migration. Control aortography confirmed good stent position, covering the entire length of DA with-

out protrusion into the aorta and with mild protrusion in the lumen of the left PA, without the risk of perforation. The flow through the stented DA was considerably higher, with a significant expansion of the entire left pulmonary vasculature (Figure 4). Arterial oximetry recorded a significant increase of the arterial saturation of 21% (from the SatO₂ of 55% before the intervention to 76% afterwards). Following the intervention, the infant had a 24-hour, continuous infusion of heparin at a dose of 20 IU/kg/hour, and commenced anti-aggregation therapy with aspirin.

After the interventional procedure, echocardiography confirmed plentiful, continuous flow through the stented DA, with a maximum pressure gradient of 46 mmHg, and a significant increase in diameter of the left PA. Further

clinical follow-ups verified an improvement in the child's growth-progress and satisfactory arterial SatO₂, with a trend of slow gradual reduction over time. One year after DA stenting, heart catheterization was repeated, in which the SatO₂ was 79%, with significant in-stent stenosis of DA (narrowest diameter: 1 mm) and excellent development of the left PA (diameter: 9 mm).

DISCUSSION

Stenting of DA represents an extraordinary technical challenge in duct-dependent CHD, since the procedure is generally performed where DA is the only source of pulmonary or systemic circulation. In these critical hemodynamic situations, even the smallest mistake or inexperience can lead to fatal consequences [2-5]. Common additional problems in the correction of duct-dependent CHD are variable morphology and spatial orientation of DA, which make the stenting procedure even more challenging. The anatomy and the spatial pattern of DA determined the optimal vascular access (femoral, axillary, carotid or trans-pulmonary) and characteristics of the stent. When choosing a stent, special attention should be paid to the length, diameter, and stent design. A stent with a larger diameter, made of thick mesh with smaller holes, allows better support of the DA wall and tends less towards causing prolapsing intraluminal tissue and consequent in-stent restenosis. On the other hand, this feature decreases the stent flexibility, which is very highly recommended in case of tortuous forms of DA. Special attention must be paid to fully covering the pulmonary end of DA with the stent, as this area has the greatest potential for constriction to the point of complete closure. Therefore, one of the most important aspects of the procedure is choosing the correct length of the stent in order to cover the entire length of DA,

without protrusion into the aorta and with minimal protrusion into the PA, thus preventing constrictive reactivity of the DA terminal part.

Since the stented DA is physiologically similar to the central aortopulmonary shunt, an optimal stent diameter is 3-4 mm for neonates, because the excessive expansion of DA may produce unwanted pulmonary overflow. Another potential problem in the aftermath of the procedure is tissue proliferation with subsequent neointimal "in stent" stenosis and that is why children should receive continuous antiplatelet therapy. In the case of early in-stent stenosis and a significant drop of arterial saturation, it is possible to perform re-dilatation or even re-stenting of the arterial duct.

Stenting of DA, in comparison with the classical surgical aortopulmonary (AP) shunt, has many advantages: lower physical trauma, avoidance of thoracotomy and the formation of adhesions, faster recovery, and less expensive treatment [1]. However, durability of the stent in DA is limited with neointimal proliferation and potential thrombosis [6]. Therefore, attempts have been made to design new antiplatelet drugs or drug eluting stents that will solve these problems [7, 8]. Furthermore, there are several common complications in relation to DA stenting, including the vascular injury of entry vessels, perforation of DA or pulmonary artery, inadequate position or dislocation of the stent and worsening branch pulmonary artery stenosis [9].

In summary, the tendency of faster evolution towards the obstruction of a stent means the surgical AP shunt is no more imperfect than the non-surgical method, as both procedures are palliative and limited in duration until the appropriate age and growth of the pulmonary artery is reached [10, 11]. Considering everything, despite the existence of some technical and clinical limitations, stenting of DA can be an effective alternative to primary surgical correction in selected patients with the duct-dependent CHD.

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Транскатетерско стентирање отвореног артеријског дуктуса код дуктус-зависних урођених срчаних мана

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КРАТАК САДРЖАЈ

Увод Најкритичније урођене срчане мане су зависне од артеријског дуктуса (*ductus arteriosus* – *DA*) и захтевају стабилну комуникацију системског и плућног крвотока. У почетном току се даје простагландин Е1 за постизање сталне отворености *DA*, а затим се хируршки креира неки од аортопулмоналних шантова. Због ризика и компликација хируршких захвата код новорођенчади с критичним урођеним срчаним манама, у новије време се алтернативно у одабраним случајевима може покушати транскатетерско стентирање *DA*.

Приказ болесника Четворомесечном одојчету дијагностикована је комплексна цијаногена урођена срчана мана типа атрезије плућне артерије с вентрикуларним септалним дефектом и искључивим снабдевањем левог хемиторакса из *DA*, док се десни хемиторакс напајао са три аортопулмоналне колатерале. Транскатетерски је без компликација

постављен коронарни стент у критично стенотични и дугачки *DA* с крајњом димензијом лумена стента од 3,5 *mm* и значајно повећаним плућним протоком лево. После интервенције дете је знатно боље напредовало, а оксиметријски је забележен значајан скок артеријске засићености кисеоником (21%). Контролном аортографијом годину дана после постављања стента утврђена је стеноза *in-stent* са значајно повећаним пречником леве плућне артерије и њених грана, након чега је конзилијарно одлучено да су се стекли услови за даље кардиохируршке корекције.

Закључак Стентирање *DA* је у одабраним случајевима ефикасна алтернатива хируршким методама у примарној корекцији дуктус-зависних урођених срчаних мана.

Кључне речи: урођене срчане мане; *ductus arteriosus* зависне; транскатетерска интервенциона процедура; стентирање *ductus arteriosus*

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