CASE REPORT

Genesis stent implantation without using a long sheath in two children

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Summary A Genesis stent was implanted in two children, one with superior vena caval (SVC) stenosis and one with pulmonary artery branch stenosis. Case 1 was a 2-month-old baby with SVC stenosis following intracardiac repair for total anomalous pulmonary venous connection (TAPVC) and case 2 was a 2-year-old child with left lower pulmonary artery stenosis following one-stage unifocalization for dextrocardia, double outlet right ventricle, ventricular septal defect, pulmonary atresia and major aortopulmonary collateral arteries. Both procedures resulted in immediate clinical and hemodynamic improvement. The Genesis stent has a closed-cell design with sigma hinges interpositioned between each cell. With improved deliverability and expandability of the stent, we can easily deliver it through smaller sheaths, which will facilitate its use in infants and smaller children with vascular stenosis. © 2008 Japanese College of Cardiology. Published by Elsevier Ireland Ltd. All rights reserved.

Case report

The Genesis stent (Cordis, Warren, NJ, USA), which was initially developed for the treatment of biliary stenosis, is very flexible, can traverse tortuous vessels, and has good radial strength with a lower profile delivery system [1,2]. There are some reports of the advantageous properties of the Genesis stent and its potential use in infants and small children with congenital heart disease. We attempted implantation of the Genesis stent without using a long sheath in a 2400-g baby with superior vena...
caval (SVC) syndrome complicating repair of total anomalous pulmonary venous connection, and in a 2-year-old child with left pulmonary artery stenosis after percutaneous transluminal angioplasty (PTA) complicated by intimal dissection.

Written informed consent for stent implantation and off-label use of the Genesis stent was obtained from the parents of both patients. The ethical committee of our hospital also approved its use.

Case 1

A 2-month-old baby weighing 2400 g developed SVC syndrome 40 days after intracardiac repair of total anomalous pulmonary venous connection (TAPVC) at 5 days of age. Angiography 50 days after surgery demonstrated a tight stenosis of the SVC with a pressure gradient of 11 mmHg (Fig. 1) and mild left pulmonary venous obstruction. The SVC syndrome was progressive and was complicated by persistent chylothorax, unresponsive to 2 weeks of intensive medical treatment, while the pulmonary venous obstruction was tolerated. Therefore, we planned stent implantation for the SVC stenosis 54 days after surgery. The procedure was performed under general anesthesia with orotracheal intubation. A 5F sheath was placed in the right femoral vein and 100 U/kg of heparin was administered. Without using a long sheath, a Genesis stent on a Slalom with a diameter of 5 mm and a length of 15 mm (PG1550BSS) was advanced into the SVC and was deployed in the area of stenosis near the SVC right atrium (RA) junction which was confirmed by a test angiogram from the left arm. The post-stent angiogram revealed excellent flow through the SVC with no evidence of narrowing (Fig. 2). There was no pressure gradient across the stenosis. He was heparinized overnight with an intravenous infusion of 200 U/(kg·day). The post-produce antithrombotic regimen consisted of oral dipyridamole 3 mg/(kg·day) and warfarin 0.02 mg/(kg·day). Although he died of pulmonary venous occlusion 1 month after stent implantation, there was no recurrence of the SVC obstruction.

Case 2

A 2-year-old child with 47XYY karyotype, dextrocardia, double outlet right ventricle, ventricular septal defect, pulmonary atresia, and major aortopulmonary collateral arteries underwent midline...
one-stage complete unifocalization at 4 months of age and intracardiac repair at 1 year of age. His body weight was 10.8 kg and height was 86.7 cm. He had PTA for left lower pulmonary artery stenosis at 13 months of age, which was complicated by intimal flap formation. Stenosis persisted around the previous intimal flap. Re-PTA at 1 year 8 months of age was ineffective. A pulmonary perfusion scan revealed severe hypoperfusion of the left lower lung. At cardiac catheterization under general anesthesia with orotracheal intubation, the main, left and right pulmonary artery pressure were 38/6 (mean, 20), 38/6 (20), and 26/6 (15) mmHg, respectively. A left pulmonary angiogram demonstrated stenosis of the left lower pulmonary artery (Fig. 3). We decided to implant a Genesis stent to rescue it. A 6F sheath was placed in the right femoral vein and 100 U/kg of heparin was administered. A 6F long sheath (Brite tip, Cordis) was positioned in the main pulmonary artery. After predilation using a SASUGA (Boston Scientific, Natick, MA, USA), with a balloon diameter of 5 mm and a length of 2 mm, the Genesis stent on a Slalom with a diameter of 5 mm and a length of 15 mm (PG1550BSS) was deployed in the lesion. Although we introduced the long sheath in the main pulmonary artery, it was not advanced through the lesion because of tortuous and stenotic blood vessels. Consequently, the stent was advanced to the lesion from the long sheath in the main pulmonary artery. The poststent angiogram revealed excellent flow through the lesion (Fig. 4). He was heparinized overnight with an intravenous infusion of 200 U/(kg day). The post-procedure antithrombotic regimen consisted of oral ticlopidine HCl 5 mg/(kg day) and aspirin 2 mg/(kg day).

**Discussion**

In Japan, intravascular stenting has been increasingly used for various congenital and postoperative vascular stenoses even in children. Although various new generation stents have been released in North America and Europe, the only stent approved for use in large vessels in Japan is the original Palmaz. It has several limitations when used in infants and small children, including a rigid strut, a sharp edge, and the need for a relatively large sheath to deploy it [2]. Even a medium sized original Palmaz stent needs a 7F sheath to deploy, while the larger Palmaz requires an 8F or larger sheath depending on the size of the balloon on which the stent is mounted.

There are a few reports on stenting for SVC obstruction in infants [3]. Frias et al. [4] reported...
the successful implantation of a P188 stent in a 3.5 kg infant with SVC stenosis using an 8F long sheath. Case 1 in the present study required relief of the SVC obstruction to avoid cerebral hemorrhage, infection, and hypo-nutrition following the persistent drainage of chyle from the right pleural cavity and high venous pressure despite intensive medical management. Although balloon dilation is one management option, immediate restenosis can occur due to elastic recoil after the procedure [5]. Considering it, we decided to implant a stent. Although the medium sized Genesis stent, which can go through a 5F sheath unlike a medium or large original Palmaz, has limited re-dilatation potential compared to the original large Palmaz, because of the low weight of our baby, we chose the Genesis stent.

In case 2, stenting of pulmonary artery branch stenosis may contribute to reducing pulmonary regurgitation and preserving as much pulmonary vasculature as possible [6]. Considering the small area supplied by the left lower pulmonary artery, we chose to implant a medium sized Genesis stent. We placed a 6F long sheath in the main pulmonary artery, because it was difficult to deliver it through the left lower pulmonary artery. However, the Genesis stent could be easily delivered to the left lower lesion without the use of a long sheath. A long sheath is sometimes difficult to position, and its application may be associated with vascular complications or hemodynamic compromise, especially in infants and small children [7]. Our experience suggests that the Genesis stent can be implanted in SVC and pulmonary branch stenosis without using a long sheath even in infants and small children with congenital heart disease. Early introduction of the Genesis Extra Diameter series, which has a potential maximal diameter of 18 mm, and is available in lengths from 19 to 59 mm, is warranted in our country.

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References