Mitral implantation of a (Melody) pulmonary valve. A report of the first pediatric case in Colombia

Implante de válvula pulmonar (Melody) en posición mitral. Reporte de primer caso pediátrico en Colombia

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Abstract

Congenital mitral valve disease is complex, entails a significant challenge from a clinical, hemodynamic, and surgical point of view, and has high rates of morbidity and mortality. Surgical valvuloplasty has a high failure and reoperation rate, especially in neonates and infants due to the presence of dysplastic valves and special anatomical characteristics. Mitral valve replacement presents limitations in this age group given the lack of available prostheses for rings smaller than 15 mm. Melody valve implantation in mitral position is a management alternative in these patients with important potential benefits in the short- and medium-term. We present the case of a 6-month-old girl with a congenital heart disease consisting in severe mitral valve stenosis, severe aortic coarctation and hypoplasia of the transverse aorta and the isthmus. She was taken to surgery at 4-months of age for a reconstruction of the aortic arch, mitral valvuloplasty, and creation of a calibrated 5 – mm interatrial communication. She developed early failure of the valvuloplasty with stenosis and severe residual mitral regurgitation, and progressed to severe acute heart failure. It was decided to implant a pulmonary Medtronic Melody 22 mm valve in the mitral position by a hybrid approach in collaboration with the pediatric hemodynamics group and cardiovascular surgery.

Keywords: Congenital heart disease. Mitral valve. Prostheses and implants. Pediatrics.

Resumen

La enfermedad congénita de la válvula mitral es compleja, implica un reto importante desde el punto de vista clínico, hemodinámico y quirúrgico, y tiene altos índices de morbilidad y mortalidad. La valvuloplastía quirúrgica tiene una tasa alta de fracaso y reintervención, especialmente en los recién nacidos y lactantes, dada por la presencia de válvulas displásicas y sus características anatómicas específicas. El reemplazo de la válvula mitral tiene limitaciones en este grupo etario por la falta de prótesis disponibles para anillos con diámetros menores a 15 mm. La implantación de una válvula Melody en la posición mitral es una alternativa de manejo en estos pacientes con potenciales beneficios importantes al corto y mediano plazo. Presentamos el caso de una niña de seis meses de edad con cardiopatía congénita dada por estenosis mitral severa, coarctación de aorta severa, e hipoplasia de la aorta transversa y el istmo. Fue llevada a cirugía a los cuatro meses de edad para una reconstrucción del arco aórtico, valvuloplastia mitral y la creación de una comunicación interauricular calibrada de 5 mm.
Presentó falla temprana de la valvuloplastia con estenosis e insuficiencia mitral residual severa, y evolucionó a falla cardíaca severa. Se decidió realizar un implante de una válvula Medtronic Melody pulmonar de 22 mm en la posición mitral a través de un abordaje híbrido en colaboración con el grupo de hemodinamia pediátrica y cirugía cardiovascular.


Introduction

Congenital mitral valve disease is complex and associated with other heart diseases, generally obstructive lesions of the left heart\(^1\). Medical treatment and surgical valvuloplasty remain a challenge in children under 2 years of age with high morbidity and mortality\(^2\). Surgical valvuloplasty has a high failure and reintervention rate, especially in neonates and infants, due to the presence of dysplastic valves and special anatomical characteristics\(^3\). Mitral valve replacement presents limitations in this age group, given the lack of available prostheses for rings smaller than 15 mm\(^4\).

The Medtronic Melody valve is a stent–mounted bovine jugular vein graft approved for percutaneous implantation for the right ventricular outflow valve dysfunction as it has shown to restore competence and relieves obstruction of the ventricular tract\(^5\). As a result of these achievements, the Melody valve was adapted for ativoventricular valve replacement showing favorable short- and medium-term outcomes for the treatment of dysfunctional mitral valves in small cohorts of patients\(^6\)–\(^8\) by demonstrating that it can be expanded in the catheterization laboratory after somatic growth, thus avoiding early surgical reintervention\(^9\).

We present the case of a 6-month-old girl with severe mitral valve stenosis and severe aortic coarctation that was taken to surgery at 4 months of age for aortic arch repair and mitral plasty. She developed severe acute heart failure in the postoperative due to early failure of the mitral plasty, a double lesion of the mitral valve, stenosis, and severe regurgitation, for which a Melody valve was implanted in mitral position.

Clinical case

A 6-year-old girl, with a weight of 5.4 kg and height of 62 cm, with congenital heart disease consisting of severe mitral valve stenosis, severe aortic coarctation, and hypoplasia of the transverse aorta and the isthmus. She was taken to surgery at 4 months of age for a reconstruction of the aortic arch, mitral valvuloplasty and creation of a 5–mm calibrated interatrial communication. In the first postoperative week, she progressively evolved to severe acute heart failure despite maximum drug therapy and required to stay in the intensive care unit with dependence on an inodilator in continuous infusion (milrinone). The echocardiogram showed a double mitral lesion with stenosis, severe regurgitation, and biventricular dysfunction (Fig. 1A–C).

Taking into account the size limitations of the prosthesis available for the patient and the poor availability of donors for heart transplantation in this age group, it was decided to implant a pulmonary valve, in this case the 22 mm Medtronic Melody valve, in the mitral position by a hybrid approach in collaboration with the pediatric hemodynamics group and cardiovascular surgery.

Description of the procedure

After obtaining informed consent, the procedure was performed in the operating room with extracorporeal circulation and cardiopelia.

The approach was made through the left atrium. A severely diseased, perforated mitral valve with a reduced annulus was seen (Fig. 2A). The anterior and partially the posterior leaflet were resected to free the annulus, allowing the passage of an 18 mm spark plug. A 15 mm diameter polytetrafluoroethylene (PTFE) tube graft was anastomosed to the mitral annulus in its anterior aspect and partially to the posterior wall of the left atrium (Fig. 2B).

The pediatric hemodynamics group prepared the valve and verified its sufficiency. Subsequently, the struts at both ends of the stent were bent outward, leaving the three struts that support the valve commissures intact and reducing the length of the stent to 15 mm (Fig. 2C). The valve was mounted on an 18 mm × 20 mm XXL TM Esophageal balloon (Boston Scientific) and was implanted in the mitral position, being impacted on the PTFE tube and subsequently expanded to 18 mm in diameter (Fig. 2D). Its sufficiency was verified, showing that it was normally functioning (Fig. 2E). A transesophageal echocardiography showed the Melody valve in mitral position, without signs of stenosis or insufficiency, with a mean gradient of 2 mm Hg and no obstruction of the left ventricular outflow tract.
The patient had a favorable evolution. On the fifth post-operative day, a transthoracic echocardiography was performed, which showed the Melody valve in mitral position with adequate function, without stenosis or insufficiency; the left ventricular outflow tract without obstruction, an atrial septal defect 4.5 mm in diameter with the left-to-right shunt, biventricular hypertrophy, and preserved function (Fig. 3A and B). Regarding the patient's clinical state, she remained with compensated heart failure and was on treatment with carvedilol 1 milligram (mg) twice a day, captopril 0.6 mg 3 times a day, hydrochlorothiazide 6 mg twice a day, and furosemide 5 mg twice a day.

At post-operative day 7, she was transferred from the intensive care unit to the hospital ward and discharged 30 days after the procedure with clopidogrel 1 mg every...
day for 3 months and acetylsalicylic acid 25 mg every day permanently.

Discussion

Despite the important advances in recent decades, the treatment of congenital mitral valve disease entails a significant challenge from a clinical, hemodynamic, and surgical standpoint\textsuperscript{4,10}. In the pediatric population, valve surgical repair is feasible and preferable in most cases, however, in patients with irreparable defects, valve replacement is the only option\textsuperscript{11}.

Some of the concerns regarding valve replacement are as follows: 1) the unavailability of prostheses smaller than 15 mm in diameter; 2) fixed-sized prostheses do not adapt to somatic growth and the earlier the replacement is made, the earlier it will require surgical reintervention to increase the size of the valve\textsuperscript{5,12}; and 3) while bioprostheses tend to degenerate rapidly in young patients, mechanical prostheses require anticoagulation, which is difficult to regulate in children\textsuperscript{12,13}.

Studies with short- and medium-term follow-up indicate that the implantation of the Melody valve in mitral position in irreparable lesions with rings smaller than 15 mm in diameter has promising results with a high success rate. It improves both stenosis and valve insufficiency and the procedure allows the possibility of effective balloon expansion by a percutaneous approach, according to the patient’s somatic growth, which delays the need for future surgeries\textsuperscript{9,14}. Advances both in the technique and in the design of the valve, specifically for surgical implantation, will reduce in the future the inconveniences presented to date, such as the structural deterioration of the valve secondary to paravalvular leakage, central regurgitation due to perforation of the valve or no coaptation, and endocarditis\textsuperscript{8,15}.

Conclusion

The evidence so far in the literature supports the implantation of the Melody valve in the mitral position as an alternative management in infants with congenital mitral valve disease and valve rings smaller than 15 mm, with significant potential benefits in the short- and medium-term.

This case report describes the successful implantation by hybrid procedure of a modified Melody valve in mitral position, in a collaboration effort between the clinical, hemodynamic, and pediatric cardiovascular surgical group in a pediatric cardiovascular referral center in Medellín, Colombia. This provides a novel alternative for high-risk infants, with the possibility of subsequent percutaneous dilations and avoiding the risks of permanent anticoagulation.

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Conflicts of interest

The authors declare that they have no conflicts of interest.
Ethical responsibilities

Protection of people and animals. The authors declare that no experiments were performed on humans or animals for this research.

Confidentiality of the data. The authors declare that they have followed the protocols of their work center on the publication of patient’s data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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