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Case Report

Cardiac Surgical Repair of Monozygotic Twins Concordant for Down's Syndrome and Complete Atrioventricular Septum Defect Case Report

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Abstract

Although the incidence of congenital heart defects in twin gestations is high, as well as in patients with Down's Syndrome (DS). The concordance of DS and congenital heart disease in monozygotic twins is very rare. Here we report the results of two 6-month-old infants, monozygotic twins, concordant for DS and Complete Atrioventricular Septum Defect (c-AVSD) undergoing cardiac surgery. Surgical repair was scheduled for both patients during concomitant hospitalizations. Patients were operated on consecutive days and did not present any serious complications. They were discharged on the fifth day of hospitalization using a fast track anesthesia protocol. This is the first case reporting cardiac surgical repair in monozygotic twins concordant for Down's syndrome and c-AVSD.

Keywords: Monozygotic twins; Down's syndrome; atrioventricular septal defect; Cardiac repair; Fast track surgery

Introduction

Down's syndrome (DS) or Trisomy 21 is a genetic disorder that occurs in every 500-750 live births [1]. There is a lower risk of DS in monozygotic twin gestations relative to singleton pregnancies but an increased risk of at least one twin being affected in dizygotic pregnancies [1]. To every 100 babies born with Down's syndrome, 40 are born with a heart defect, 40% of which are diseases of the atrioventricular septum [2]. Congenital heart defects (CHD), which represent the most common human birth defects, occur in approximately 8 per 1000 live births [2-4].The concordance prevalence of congenital heart diseases in monozygotic twins is around 9.5% and between 4.5-13.5% in dizygotic twins, especially if complicated by twin-to-twin transfusion syndrome [2]. Complete atrioventricular septal defect (c-AVSD) is characterized by an ostium primum atrial septal defect (ASD), a single atrioventricular (AV) valve and a ventricular septal defect (VSD), which leads to progressive heart failure, recurrent pneumonia, being usually corrected up to 6 months of life due to the risk of developing pulmonary hypertension and increased

mortality [5-7]. Despite the relatively high incidence of congenital heart defects in twin gestations and DS patients independently, the concordance of DS and CHD in monozygotic twins is uncommon. Here we describe an extremely rare case of surgical repair of the cardiac condition in monozygotic twins concordant for DS and c-AVSD using a fast track anesthesia protocol.

Case Presentation

Twin 1- a 36 weeks premature birth female infant now aged 6 months and 10 days, asymptomatic, weighing 4.7 kg, concordant for DS and Rastelli type A c-AVSD. On the morning before the surgery, managements for reducing fasting and oral premedication were accomplished. Upon arrival at the operating room, anaesthetic induction with tracheal intubation, invasive procedures, level T5 bilateral erector spinae muscles plane block with injection of 0.2% ropivacaine 0.75 ml/kg on each side were accomplished. Transesophageal echocardiogram showed an 0.5 cm Ostium Primum ASD, two 0.3 cm Ostium Secundum ASD´s, 1 cm Inlet VSD, single AV valve with mild left and moderate right AV regurgitation by the cleft and no signs of pulmonary hypertension (Figure 1). Surgical repair consisted of ventricular septoplasty, separation and repair of both AV valves with bilateral cleft closure, and atrioseptoplasty. Table 1 summarizes intraoperative variables. Total cardiopulmonary bypass time was 115 minutes with aortic clamping of 100 minutes. Post-correction transesophageal echocardiogram showed mild regurgitation of the left AV valve (Figure 2). The patient required cryoprecipitate and platelets after cardiac bypass for a clotting disorder. After the end of the surgery, the patient was extubated and sent to the ICU painlessly, lightly sedated, with infusions of dexmedetomidine and milrinone. Twin 2- a 36 weeks premature birth female infant now aged 6 months and 11 days, asymptomatic, weighing 4.8 kg, concordant for DS and Rastelli type A c-AVSD. On the morning before the surgery, the same management as for twin 1 was accomplished. After arriving at the operating room and receiving standard monitorization, the patient was submitted to inhalational anaesthetic induction while still on the mother's lap and moved to the operating table with posterior puncture of peripheral venous access, tracheal intubation, invasive procedures and level T5 bilateral erector spinae muscles plane block with ropivacaine injection, 0,2% 0.75 ml/kg on each side. Transesophageal echocardiogram showed a wide Ostium Primum ASD, Ostium Secundum ASD, a Patent Foramen Ovale, a wide inlet VSD, a single AV valve with mild left and moderate right AV regurgitation (Figure 3). Surgical repair consisted of ventricular septoplasty, separation of atrioventricular valves, left valve repair with cleft closure, right valve repair with cleft closure, atrioseptoplasty. Coincidentally, total CPB time was 115 minutes and aortic clamping time was 110 minutes. Transesophageal echocardiography after correction showed good ventricular function, mild right and left AV regurgitation (Figure 4). Awake and extubated after the end of the surgery, the patient was transferred to the ICU calm, painlessly and receiving low doses of dexmedetomidine and milrinone. Both twins had satisfactory evolutions and were discharged on the fifth day of hospital stay. Twin 1, despite having favourable conditions for discharge the day before, waited for twin 2 due to family requirement.t.

	TWIN 1	TWIN 2
By-pass/Aortic Clamp	115/110 minutes	115/110 minutes
Intraoperative Blood Products	RBC:150ml (CBP) FFP:150ml (CBP) CP:50ml PC: 50ml	RBC:120ml (CBP) FFP:120ml (CBP)
Ultrafiltration (MUF+CUF)	375ml	575ml
Fluid Balance (end of surgery)	-60ml	-135ml

RBC, Red Blood Cells; FFP, Fresh Frozen Plasma; CP, Cryoprecipitate; PC, Platelets Concentrate; MUF, Modified Ultrafiltration; CUF, Conventional Ultrafiltration; CBP, Cardiopulmonary Bypass Prime.

Table 1: Intraoperative Variables.

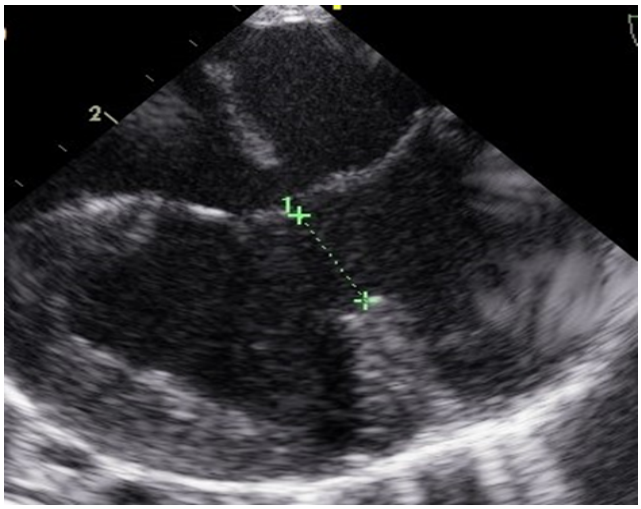

Figure 1: Transesophageal echocardiogram of twin 1 showing 0.5 cm Ostium Primum ASD, two 0.3 cm Ostium Secundum ASD´s, 1 cm Inlet VSD (dotted line) and single AV valve.

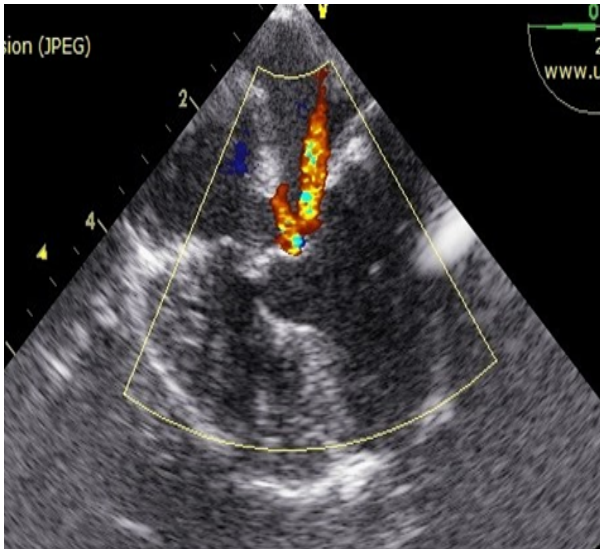

Figure 2: Post-correction transesophageal echocardiogram of twin 1 showing good surgical result, mild regurgitation of the left AV valve.

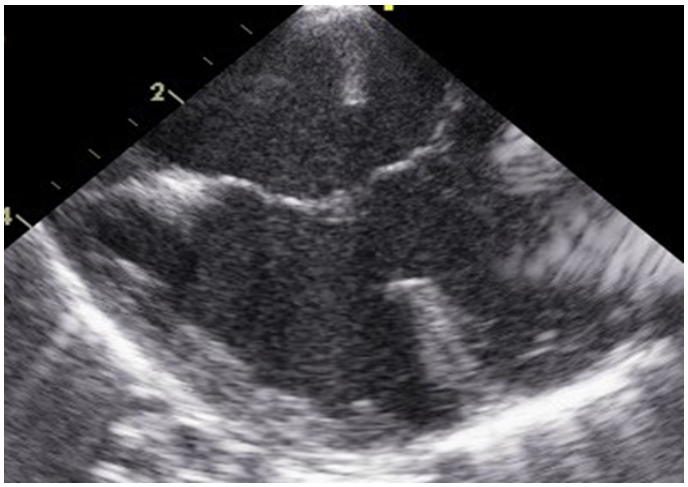

Figure 3: Transesophageal echocardiogram of twin 2 showing a wide Ostium Primum ASD, Ostium Secundum ASD, a patent foramen ovule, a wide inlet VSD and a single AV valve.

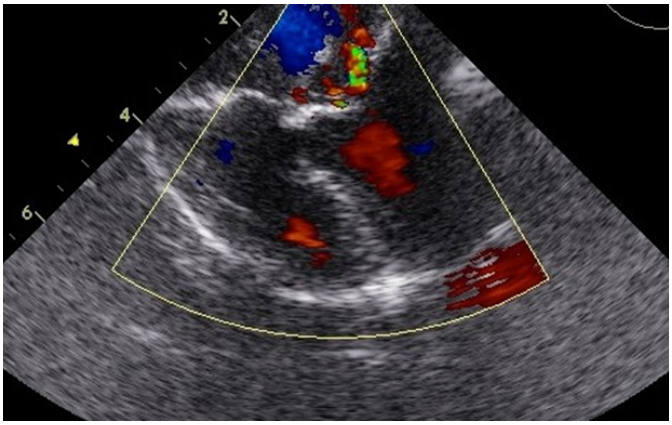

Figure 4: Post-correction transesophageal echocardiogram of twin 2 showing good surgical result, with mild right and left AV regurgitation.

Discussion

After an active search in PubMed, ScienceDirect and LILACS databases using the terms monozygotic twins, Down´s Syndrome, surgical repair, atrioventricular septal defect, in addition to other combinations in English, Spanish and Portuguese, the authors did not find references of monozygotic twins concordant for DS and c-AVSD with or without surgical correction. A case report of dizygotic (but not monozygotic) twins concordant for DS and c-AVSD showed disparate outcomes where only one had a traditional evolution and surgical repair [2]. Another report of monozygotic twins concordant for DS and congenital heart disease did not obtain an echocardiographic diagnosis, but only a report of VSD in one of the twins submitted to necropsy after death at 7 months of life. None of them underwent surgical repair [8]. Finally, in a study to determine the incidence of structural congenital heart disease in monochorionic twins, there was only one case of agreement for c-AVSD but with normal karyotypes and both died at 14 weeks of gestation due to hydrops fetalis [9]. Our report draws attention both for the rarity of DS agreement in monozygotic twins and for the absence of reports that entitle the publication. Despite the excellent surgical result that usually follows this surgery and

the initial desire of the family members to perform the procedures on the same day, we chose to perform the procedures on alternate days for safety reasons. Our protocol focused on procedures for fast track surgery, which included immediate tracheal extubation after surgery, intra and postoperative multimodal analgesia with opioid sparing techniques, delirium and nausea prophylaxis, optimization of chest tube removal, early oral intake after surgery, allowing both to be discharged together, on the fifth day of hospital stay, without complications [10,11]. The fact that they did not present pulmonary hypertension or ventricular dysfunction certainly led to a favourable outcome.

Conclusion

This is the first report worldwide of monozygotic twins concordant for DS and c-AVSD who were concomitantly submitted to surgical repair. Both patients presented satisfactory outcomes, being discharged on the fifth day of hospitalization. A fast track anesthesia protocol allowed a humanistic and rapid discharge despite the inherent comorbidities.

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