A hybrid procedure for the closure of a large muscular ventricular septal defect in a 6-month-old infant

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Case Report

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Abstract

Background: Transthoracic device closure (TTDC), also known as a Hybrid procedure, has been proposed as an alternative, less invasive approach compared to open-heart surgery for the treatment of ventricular septal defect (VSD).

Case Presentation: We present our first national case of TTDC in a 6-month-old female baby with a muscular 8mm ventricular septal defect, 3 mm atrial defect, enlarged right and left ventricle and a dilated pulmonary artery complicated by severe pulmonary hypertension.

Treatment consisted of two pulmonary artery banding attempts at the age of 2 months to control pulmonary hypertension – the interventions were combined with diuretics and angiotensin-converting enzymes inhibitors. Yet, the initial approach was suboptimal as we noticed a failure to thrive continuous sweating and tachypnea.

Because of the worsening condition at the age of 6 months, and a weight of 6.6 kg, we performed TTDC.

After median sternotomy, a 10mm muscular VSD occluder was implanted under trans-oesophagal echocardiography guidance on the beating heart. The procedure lasted 90 min and was performed without incident; the hemodynamics were stable with only a minor residual VSD. The child was extubated after 2 hours and discharged after five days from the hospital.

Conclusions: Transthoracic device closure (TTDC) is a promising treatment modality for large muscular VSD in small infants with low weight. TTDC is feasible in cases with heavy myocardial right ventricle trabeculae and who previously underwent open-heart surgery.

Full Text

First Hybrid procedure for ventricular septal defect closure in the Republic of North Macedonia

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VSD—Hybrid Procedure—Right Heart Failure—Pulmonary Artery

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Background

Ventricular septal defect (VSD) is a common congenital heart defect observed in about 30% of all cardiac abnormalities in newborns. VSD accounts for up to 40% of all congestive heart failure in infants with an incidence rate of 2 to 6 cases per 1000 live births [1].

VSD's can manifest clinically in a number of ways spanning from asymptomatic to causing growth retardation and other developmental abnormalities [2–4]. Severe symptoms are typically associated with congestive heart failure and/or the development of pulmonary hypertension [2, 3]. Transthoracic device closure (TTDC), is considered an alternative compared to open-heart surgery, to treat ventricular septal defect (VSD). The procedure is associated with the least disruptive hemodynamic consequences, avoiding cardiopulmonary bypass, and allows fast-track anaesthesia. Collectively, TTDC promotes improved patient outcomes in terms of morbidity, mortality, mechanical respiratory support, hospitalisation time, quality-of-life and cost of care [2, 3]

Pediatric cardiac surgery is still in its infancy in the Republic of Macedonia. Between 2003 and 2019 a low volume of interventions were performed at 3 clinics in Skopje; the more complex cases were sent abroad, mostly Bulgaria, Turkey and Serbia. Since early 2019, a centralised pediatric cardiac surgery program under the public healthfund was formally established in March 2019 at our clinic. To highlight the success of Macedonia's pediatric cardiac surgery program - we report here our first case of TTDC in a female baby presenting with muscular ventricular septal defect (mVSD) not amenable to surgical closure.

Case presentation

We treated a 6-month-old female baby born with multiple congenital defects; most significant was a large 8 mm muscular VSD (*Figure 1)*.. Echocardiography revealed the presence of a smaller atrial defect, right and left ventricle enlargement and the dilatation of the pulmonary artery; the child also presented with severe pulmonary hypertension. Postpartum, the baby was hospitalised with severe pneumonia and received antibiotics (Cephalosporines), Furosemide (3 mg/kg/day until the procedure), Captopril (0.5 mg/kg/day until the procedure) and Digoxin (5 µg/kg/day until the procedure). At the age of 2 months, a pulmonary artery banding was performed to control increased pulmonary flow and prevent pulmonary vascular artery disease (PVAD) as the pulmonary vascular resistance dropped. Despite the pulmonary artery banding attempt—the child's condition failed to improve—we observed clinically significant failure to thrive, tachypnea and sweating. For this reason, the child was rehospitalised (*Figure 2)*

We opted for TTDC in an attempt to mitigate the risk of intraoperative and postoperative complications; a hybrid procedure, requiring median sternotomy, to close the muscular VSD using a 10mm VSD occluder (CERA TM) on the beating heart was scheduled.

After anatomical examination of the right ventricle under transesophageal echo (TEE) guidance (10.7/8 x 27mm, Philips 7–3T), we performed a puncture with a 19 Ga needle through the right ventricle free

anterior wall, with appropriate alignment towards the interventricular septum (IVS), avoiding trabecula septomarginalis. After the tip of the needle was directed to the left side, a 0.038 wire was inserted in the left ventricle, and 7F Cera TM (curved, 180o, modified for this case) introducer was implanted, the left disc was opened, pulled towards the IVS. Next, we opened the right disc and subsequently deployed the device, without hemodynamically significant residual shunting. TTDC was performed under general anaesthesia. Inotropic support in the form of Milrinone (0.25–0.5 µg/kg/min i.v.) and Epinephrine (0.02 µg/kg/min i.v.) was administered. The total duration was 90 minutes in the absence of perioperative complications (*Figure 2 & 3)..* We continued the diuretic stimulation with Furosemide i.v (0.15–0.25 mg/kg/h for 3 days, after we switched to 3mg/kg/day). The baby was

extubated two hours postoperation on the same day with oral intake of milk formula a few hours later. The thoracic drain was removed on the first postoperative day, and the patient was discharged on the 5th postoperative day. A minor decrease in right ventricle chamber diameter (24 vs 22mm) and pulmonary annulus diameter (21 vs 19 mm) was noticed in first post interventional day. During the hospital stay, the reduction of the Right Ventricle (RV) diameter in RV-Outflow Tract (RVOT) region on long-axis parasternal view was 4 mm, and the diameter of the pulmonary annulus was decreased approximately 5 mm. The gradient in residual shunt across a small apical VSD rose from 30 mmHg to 60 mmHg during the hospitalisation. One week after discharge, follow-up examinations revealed an improving clinical state, reduction of tachypnea (24/min), and sweating and irritability. Follow up echocardiography showed a residual shunt towards the apex of the ventricle, with the colour neck of 2.5 mm, with a residual left-to-right gradient of 65 mmHg, reduction of the pulmonary artery diameter to 14.7 mm, estimated diastolic pulmonary pressure of 18 mmHg, estimated systolic pulmonary pressure of 34 mmHg, slight enlargement of the right heart (*Figure 4)..* The wound from medial sternotomy was in good condition.

Discussion

A National Congenital Cardiac Surgery program was established in 2019 in the Republic of North Macedonia. To provide the highest standard of care, we have established a capacity-building program with the Joe DiMaggio Children's pediatric cardiac surgery unit. Through the program, our local staff receives remote and on-site training and is assured of expert assistance through video conferences and ad-hoc remote supervision by the medical team in Florida.

Treatment of large hemodynamically important VSD includes surgical closure of the defect or catheterisation closure. Surgical closure is the mainstay treatment for most VSDs. Muscular VSD is challenging to access without a left ventricular incision, in particular when the VSD is accompanied by a large left-to-right shunt, pulmonary hypertension, or interventricular septal hypertrophy [1, 4]. Postoperative AV block is also more common in such cases.

The indications for treatment of large VSD in small infants are still ill-defined and are mainly guided by the individual clinical characteristics. Failure to thrive, congestive heart failure, a refractory medical condition, worsening pulmonary hypertension are the most critical signs of large VSD and indicators for closure [2, 5]. The patient's age and weight should also be considered in the decision-making process.

In infants, large muscular VSD is challenging to close from the right ventricular side. Pulmonary artery banding initially to control CHF and reduce the pulmonary artery pressures is performed in young (up to three months of age) babies. Open surgical repair is associated with the risk of effects of cardiopulmonary bypass (CPB), the risk of left ventriculotomy in large mVSD because of an inadequate operative field of vision, and postoperative ventricular dysfunction and arrhythmias. The estimated mortality rate following open-heart surgery ranges between 3 - 8% [6]

Nevertheless, hybrid procedures, such as transthoracic device closures (TTDC) pose less risk than conventional surgical closure of the muscular VSD. Per-ventricular device closure of VSD with TEE guidance was first reported by Amin and colleagues [7]. The method combines surgical technique and interventional closure and makes it possible to avoid possible complications. The so-called hybrid approach of device closure of muscular ventricular septal defect has been encouraging and has been used in a few centres. This novel approach can be used to avoid not only cardiopulmonary bypass but also vascular injures caused by interventional closure.

Accounting for the factors mentioned above, our patient's age, weight, clinical state, developing pulmonary hypertension despite previous pulmonary banding, the dimension of the VSD and localisation we opted for the TTDC approach. The anatomical aspect is crucial, but also our goal to avoid left ventriculotomy. Furthermore, TTDC proceeds without extracorporeal circulation and thus may lower the risk of postprocedural atrioventricular blocks (AVB).

A recent meta-analysis by Yang Zhou et al., [8] supported the decision for TTDC. The analysis included a total of 5 randomised controlled trials, 7 cohort studies, 13 case-control studies, 129 case series and 13 case reports. The primary outcome of interest was the success rate; the proportion of patients with no residual shunts, intraoperative conversion to open-heart surgery and morbidity from perioperative complications, for example, arrhythmia, valve regurgitation (aorta, tricuspid valve), AVB, as well as duration of the procedure, intensive care stay, hospital stay, number of transfusions. According to this study, we do not find intraoperative arrhythmia, or new onset of aortic or tricuspid regurgitation, due to device proximity toward the valves. The authors concluded that there was a significant difference in the rates of residual shunts comparing TTDC and open-heart surgery. Risk factors associated with for postoperative AVB were the existence of large muscular VSD, female gender and the size of the occluder.

Despite the presence of several risk-factors, we observed an uncomplicated postoperative course in our first successful case of TTDC which was further characterised by fast extubation, same-day oral feeding, mother-daughter bonding and discharge within one week.

The duration of the treatment of 90 minutes (*Figure 3)* can be explained considering the precautious measures taken, among others, a median sternotomy was done in the case we had to convert to open-heart surgery. Additionally, we modified the 7F Cera introducer for this purpose, to allow better control and opening of the device. However, despite the relative longer procedure time, there was no bleeding, need for intraoperative transfusions, occluder fall, malposition or malalignment of the device towards the intraventricular septum.

Limitations

This work describes an initial case report. The long-term prognosis should be carefully evaluated in light of the residual shunt. We successfully reduced the dimensions of VSD to a level that is acceptable for the age and weight and which should promote complete closure with time. However, on the first postoperative control one week after discharge, even though we observed a markedly improved clinical state, there was still a residual < 2 mm shunt. To this end, intensive follow up will be required and likely more aggressive medical therapy (diuretics, ACE blockers).

Conclusion

Transthoracic device closure (TTDC) is a promising non-invasive modality for large muscular VSD in small infants, even in a child with low body weight. TTDC is feasible in the presence of significant myocardial right ventricle trabeculae and following previous open-heart surgery.

Figures

List of abbreviations

AVB = Atrial ventricular Block

TTDC = Transthoracic Device Closure

TEE = Transesophageal Closure

VSD = Ventricular Septal Defect

Declarations

Ethics approval and consent to participate

The ethical committee approved the clinical practice and treatment procedures described.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images; the use of all health and medical information for scientific research and manuscript preparation was approved. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Availability of data and material

All original data described in this case report can be submitted for evaluation upon request

Competing interests

Dr Zan Mitrev is the hospital director at the Zan Mitrev Clinic.

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Not applicable; no funding was received for this case report.

Authors' contributions

IM and ZM performed the procedure. DP was responsible for anaesthesia and perioperative care. DP, IM and RR wrote the manuscript. All authors have read and approved the manuscript. SB provided clinical assistance, expertise and critically reviewed the manuscript

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None

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& Figure Legends

Figure 1—Echocardiography evaluation a 6-month-female baby with the ventricular septal defect; Apical 4-Chamber view on TEE (56)*

The first panel shows an enlarged left and right ventricle with a mid-portion significant muscular ventricular septal defect (*A)* A left-to-right shunt through muscular VSD (*B)* Measurement of the right ventricle to obtain an appropriate length of the introducer in the left side (*C)* The left disc of muscular VSD occluder is open in the left ventricle; measurements of the distance toward to right side (*D);*; the left disc is open with proper alignment towards interventricular septum (*E);*; both discs of the device are opened with adequate alignment towards septum (*G);*; both discs of the device are opened, with proper alignment towards septum, measurement of the right ventricle below the tricuspid valve; (*H)* Apical 4 Ch view, the position of the occluder (left side) colour Doppler left to right shunt (right side); (*I)* preoperative left-to-right shunt and (*J)* preoperative pulmonary annulus.

Figure 2—Digital tracking of a 6-month-old female admitted with pulmonary hypertension resulting from 10mm ventricular septal defect.

00*The p*atient was admitted on the 26th of May 2020 () and underwent a hybrid procedure on the next day (). The temperature, weight, diuresis, and biochemistry analyses are shown as automatically documented using digital electronic health record.

Figure 3—Intraoperative and postoperative digital tracking of Blood Gas Analysis values of a 6-month-old female who underwent a hybrid procedure to manage a 10mm ventricular septal defect.

00(*B)* The patient was placed under general anesthesia on the 27th of May, 07:25 until 10:40 (). A median sternotomy was done at 09:05 and closure at 10:15. Analysis of blood gas values was initiated 07:54 and repeated during the postoperative course (*B)* HCT = Hematocrit (%), HGB = Hemoglobulin (g/dL), K = potassium, N = Sodium, MetHb = Methemoglobin, COHb = Carboxyhemoglobin (COHb), O2Hb = oxygen-carrying hemoglobin, Ca = calcium, pCO2 = partial pressure of carbon dioxide, pO2 = partial pressure of oxygen

Figure 4—Follow-up control echocardiography evaluation; 4 weeks after a hybrid procedure to close a muscular VSD

Panel (*A)* shows the parasternal axis view of the Left Ventricle, Intraventricular septum and Right Ventricle and a minor left to right shunt with a gradient of 85 mmHg. (*B)* The long parasternal axis view of the Left Ventricle; the apical part of the IVS, with residual VSD and occluder in mid-portion. (*C)* short parasternal axis view, showing the diameter of pulmonary annulus after 3 weeks (15mm). (*D)* subcostal axis view, residual ASD fossa ovalis type, 3mm, spontaneous left to right shunt. (*E)* Long parasternal axis view, the diameter of the right ventricular outflow tract. (*F)* Five-chamber apical view and transaortic flow. (*G)* Long parasternal axis view showing M-mode measurements of the Left Ventricle.

Supplementary Files

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