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Case Report

Stenting the Ductus Arteriosus of a Patient with Tetralogy of Fallot and Trisomy 18: Case Report and Updated Review

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Abstract: Ductal stent implantation is a palliative procedure that can be performed in cases of congenital heart defects dependent on the patency of the ductus arteriosus, such as Tetralogy of Fallot. It is a widely effective technique, with low complication rates and less invasive than the modified Blalock-Taussig palliative surgery, making it a viable and safe option for high-surgical-risk patients. Moreover, it is crucial for the anesthesiologist to be familiar with the pathophysiological characteristics of these ductus arteriosus-dependent heart conditions to ensure appropriate perioperative management. The objective of this work is to describe the challenging anesthetic management of a newborn with Tetralogy of Fallot and Edwards Syndrome who underwent ductal stent implantation via endovascular approach at a quaternary care center.

Keywords: Tetralogy de Fallot; Ductus Arteriosus; Patent Ductus Arteriosus.

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1. Introduction

Congenital heart diseases with duct-dependent pulmonary circulation, such as Tetralogy of Fallot (TOF), represent a group of life-threatening conditions. Maintenance of pulmonary blood flow through the ductus arteriosus is essential for the survival of patients affected by these conditions [1, 2]. TOF affects approximately 1 in every 3,500 births and accounts for 7-10% of all congenital heart diseases. These patients may experience severe hypercyanotic episodes triggered by factors such excessive stimulation of the sympathetic nervous system caused by pain or anxiety, hypoxia, hypercarbia, acidosis, and the induction of general anesthesia [1, 3, 4].

Several palliative therapeutic strategies are available, such as prostaglandin infusion and palliative surgical procedures. Given the high morbidity and mortality associated with the modified Blalock-Taussig shunt, an equally effective and less invasive technique has been described to ensure proper pulmonary blood flow, known as patent ductus arteriosus stenting [5, 6]. This study aims to report on the case of a term newborn with TOF associated with trisomy 18, who underwent endovascular ductal stent implantation as a palliative procedure, as well as to describe the particularities of anesthetic management and the surgical procedure.

2. Case Report

Thirty-eight weeks full-term newborn, weighing 2,035g (0,5th percentile), measuring 42cm (0,5th percentile), whose prenatal fetal karyotype detected trisomy 18. Born by cesarean section due to fetal malformation, the newborn received the initial steps of neonatal

resuscitation and was transferred to the intensive care unit. A transthoracic echocardiogram was performed on the day of birth, diagnosing TOF with following unfavorable abnormalities: right ventricular outflow tract obstruction, ventricular septal defect, overriding aorta and hypoplastic pulmonary arteries. The main echocardiographic findings are summarized below (Table 1). The diameter of the ductus arteriosus was not measured. Continuous prostaglandin infusion was initiated.

Table 1. Main initial echocardiographic findings.

1	Patent foramen ovale (Left-to-Right)
2	Right atrial dilation and right ventricular hypertrophy, with significant right ventricular outflow tract obstruction
3	Tricuspid valve: dysplastic, antegrade flow and mild regurgitation
4	Perimembranous ventricular septal defect with right-to-left shunt
5	Aortic valve: annulus involves 50% of the interventricular septum; dysplastic valve with dome-shaped opening and turbulent flow; mild regurgitation
6	Pulmonary valve: dysplastic, with thickened leaflets and absence of antegrade flow
7	Main pulmonary artery: significant reduction in diameter, with confluent and mildly hypoplastic pulmonary branches (Left PA: 2.7 mm / Right PA: 2.6 mm)
8	Patent ductus arteriosus: small, tortuous, with left-to-right shunt (Ao-to-PA)
9	The right and left coronary arteries appear normal

The pediatrics team informs about the possibility of stent implantation in the ductus arteriosus as a palliative approach, depending on the care plan to be discussed with the family. A meeting was held with family members, explaining the existing therapeutic options and the risks inherent to each technique. After consent and authorization from the family, the decision was made to proceed with ductus arteriosus stent implantation. The patient was received in the hemodynamic room under basic monitoring. The continuous prostaglandin infusion was stopped six hours before the procedure. Anesthetic induction was performed with fentanyl, ketamine, and rocuronium. Orotracheal intubation was carried out under direct laryngoscopy. Anesthetic plan maintained with sevoflurane.

Heparin was administered and the activated clotting time was measured at 380 seconds. The left axillary artery was punctured, and a 4F introducer was inserted (Figure 1). Pulmonary artery catheterization was performed, followed by ductus arteriosus stent implantation (Figure 2). During the procedure, the patient experienced periods of significant desaturation, prompting the administration of a crystalloid bolus and the initiation of continuous phenylephrine infusion. At the end of the surgery, the patient was transferred to the intensive care unit. In the first hour post-operatively, the patient developed persistent pulse oximetry drop and central cyanosis. The prostaglandin infusion was restarted.

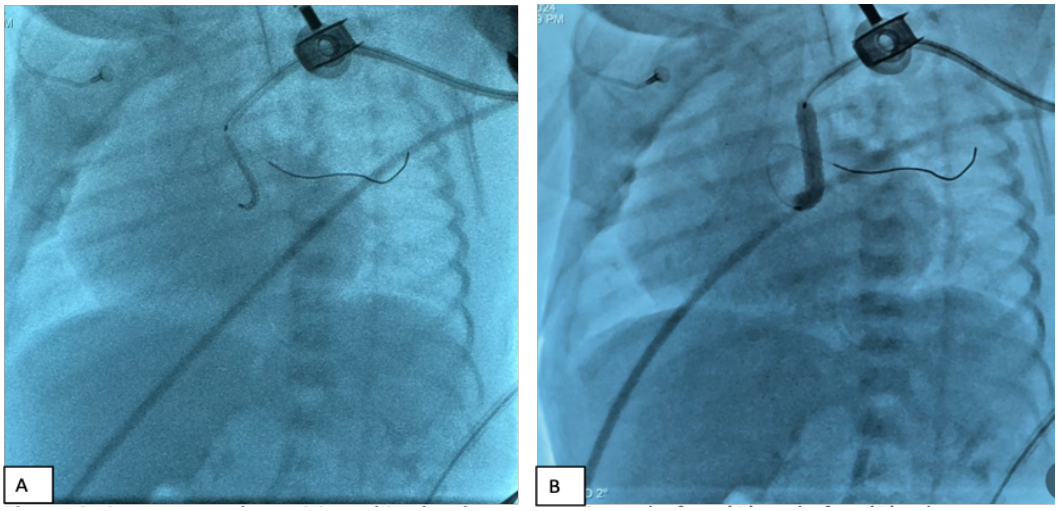
A new transthoracic echocardiogram was performed on the day following the procedure and revealed a ductal stent with suboptimal positioning. Both the aortic and pulmonary ends of the ductus arteriosus remained patent, but the aortic end was not fully covered by the stent (in-stent pressure gradient of 39 mmHg). As the patient's clinical condition deteriorated during the immediate postoperative period, invasive ventilatory and hemodynamic management became necessary and required frequent adjustments, precluding early extubation and weaning from the vasoactive agents and prostaglandin. The care plan, discussed by the pediatric intensivists with the family, prioritized comfort measures, and together with the family, it was decided not to pursue further therapeutic interventions given the severity of the case, the guarded prognosis, and the unfavorable outcomes associated with additional therapeutic attempts.

In the following days, required high mechanical ventilation settings and infusion of high doses of phenylephrine and prostaglandin. A new echocardiogram was scheduled for the 6th postoperative day, but on the 5th postoperative day, the patient experienced a

cardiorespiratory arrest and passed away, with inadvertent displacement of the implanted stent being one of the possible diagnostic hypotheses. In fact, the infectious laboratory review was unremarkable.

Figure 1. Percutaneus vascular access of the left axillary artery with a 4F introducer sheath.

Figure 2. Stent properly positioned in the ductus arteriosus before (A) and after (B) release.



3. Discussion and Conclusion

Cyanotic congenital heart defects dependent on the ductus arteriosus may necessitate palliative surgical procedures until the patient is stable enough to undergo definitive repair [6]. TOF is a cyanotic congenital heart defect characterized by a ventricular septal defect, right ventricular outflow tract obstruction, overriding aorta, and right ventricular hypertrophy. The outflow tract obstruction is the primary determinant of the severity of

hypoxemia [3]. For this reason, the patency of the ductus arteriosus is crucial to ensure adequate pulmonary blood flow [1]. Affected individuals may present associated chromosomal abnormalities. Trisomy 18 (Edwards Syndrome) corresponds to one of these associations and presents a poor prognosis in the first year of life, being characterized by several cardiac defects, such as ventricular septal defects, patent ductus arteriosus and polyvalvular disease [7].

Most neonates with ductal-dependent pulmonary blood flow require infusion of prostaglandin E1 to prevent premature closure of this structure. Prostaglandin E1 infusion should be stopped 6 to 12 hours prior to stenting the ductus arteriosus to facilitate the device placement in a constricted duct and prevent inadvertent stent displacement [5, 8]. Ductal stenting is performed under general anesthesia, and central venous access is imperative [5]. To prevent exacerbated sympathetic stimulation, some centers recommend the administration of pre-anesthetic medication, such as oral midazolam or intranasal ketamine. Regarding anesthetic induction, opioids are indicated for analgesia, and ketamine is the preferred hypnotic due to its ability to maintain systemic vascular resistance, thereby attenuating the risk of hypercyanotic episodes [3, 9].

Hypercyanotic episodes can be explained by the intensification of the right-to-left shunt. Therapeutic measures include reducing pulmonary vascular resistance, increasing systemic vascular resistance, and decreasing the level of right ventricular outflow tract obstruction, the reason why phenylephrine was preferred over other vasoactive drugs [3, 9]. As an alternative measure to reduce the risks of the modified Blalock-Taussig surgery, a promising palliative technique has been described, which is equally effective and less invasive. In summary, it involves implanting a metallic stent in the ductus arteriosus [2, 6]. It is considered a cost-effective technique and a viable option for high-surgical-risk patients, such as those with low birth weight (<2.5 kg), systemic comorbidities, and unusual cardiac or pulmonary artery anatomy [8]. The choice of vascular access for stent implantation is a crucial step for the success of the procedure, with the femoral, axillary, or carotid arteries being the available options [2, 5].

Among the main complications, the most notable include vascular access-related injury, stent migration, acute stent thrombosis, and ductal spasm. Ductus arteriosus with an increased diameter (≥2.5mm) is not considered suitable for stent placement due to the risk of migration and dislodgement of the device [3, 10]. However, the endovascular procedure also carries intrinsic risks, not only due to the challenging anesthetic hemodynamic management but also because of possible surgical complications. In the reported case, as inferred from the echocardiogram performed on the day following the procedure, the likelihood of stent migration is high, which explains the child's death in the subsequent days. Other hypotheses for the death were carefully examined, but, in fact, most of the laboratory review was unremarkable, while the arterial blood gas showed marked deterioration. Due to the death occurred within the hospital setting, with a hypothesis to justify it being highly suspicious, in a patient whose chromosomal pathology is known to have a guarded prognosis, the autopsy was waived as it would not have the potential to change the care provided in this specific case.

Ductus arteriosus stent implantation is a less invasive palliative procedure with lower complication rates and comparable efficacy to the modified Blalock-Taussig shunt, acting as an alternative until the patient is stable enough for definitive surgery. The anesthesiologists must understand the pathophysiological peculiarities of Tetralogy of Fallot, as they play a crucial role in the perioperative, hemodynamic, and respiratory management of patients with this condition. They must also take an active role in discussions with the family, which include ethical considerations surrounding a condition with a guarded prognosis.

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Conflicts of Interest: None.

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