

FIGHTING FOR LIFE - COMPLICATED OUTCOME AFTER ARTERIAL SWITCH OPERATION

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Abstract: Transposition of Great Vessels (TGV) is a complex cardiac malformation featuring ventriculoarterial discordance. The elective surgical procedure is "arterial switch" operation, a complex intervention with survival rate 97-100%. We present a full-term child, diagnosed antenatally with TGV, who was urgently transferred, 6 days after birth, to Târgu-Mureș Emergency Institute for Cardiovascular Diseases and Transplantation. Emergency arterial switch surgery was performed. Postprocedural, re-intervention for hemostatic purpose was mandatory; afterwards, the patient developed renal failure. On postoperative day 14, the evolution was complicated by ceco-colic perforation. Further in evolution, seizures were noted along with upper caval thrombosis, right diaphragmatic paresis, and supraventricular arrhythmia. After 68 days of hospitalization and after suffering a range of complications, the patient was discharged being stable. In conclusion, acquiring the knowledge and practical experience required for the treatment of TGV leads to an increase in the survival rate.

INTRODUCTION

Transposition of great arteries is one of the most complex pathologies in pediatric cardiac surgery, being the most common cyanogen malformation. It represents 5% of all cardiac malformations, that is, 0.211 / 1000 live newborns, according to the Baltimore-Washington Infant Study.

In order to survive, there is a need for an interatrial, interventricular or great vessel "blood mixing" zone. The patency of the arterial duct is maintained with prostaglandin infusion, and in the case of a restrictive atrial septal defect, pre-balloon septostomy is performed, the so-called Raskind procedure.

The elective surgical procedure is the arterial switch, firstly described by Jatene in 1975, which represents an anatomical correction of the great arteries performed in the first few weeks of life.(1)

The intervention is performed in total cardiopulmonary bypass, in medium / deep hypothermia. Like any other surgical procedure of this magnitude, complications such as: low heart rate syndrome, arrhythmias, hemorrhage, cardiac tamponade, episodes of pulmonary hypertension, renal failure, neuro-cognitive disorders or infection may occur.(2)

CASE REPORT

A 40-week full-term female newborn, delivered via C-section, weighing 3,100 grams, with Apgar Score 8/9, was diagnosed prenatally at 29 weeks with transposition of great arteries. At 6 days after birth, she was transferred from the territory hospital on an emergency basis, for specialized treatment.

At the time of admission to Târgu-Mureș Emergency Institute for Cardiovascular Diseases and Transplantation, the general status of the newborn was severely impaired, respiratory

assisted, fed through a nasogastric tube, pale with signs of cyanosis on physical examination.

Transthoracic echocardiography performed at the time of admission confirmed the diagnosis of transposition of the great arteries with suspicion of the circumflex coronary artery, emerging from the right coronary artery.

Within this clinical-biological context, the next day, at the age of 7 days, the newborn underwent the arterial switch operation which is the "gold standard" procedure.

Surgical procedure and management

The procedure was performed (on 14th of March 2013) in total cardiopulmonary bypass after ligation of the patent ductus arteriosus, cooled at 26 degrees Celsius, with double caval cannulation and a cardioplegic solution of Custodiol administered every 90 minutes.

Regarding the position of the great vessels, the aorta was located 30° anterior to the pulmonary artery; in a standard manner the aorta was dissected at a lower level above the coronary arteries and the pulmonary artery at a higher level, under bifurcation of the pulmonary branches.

The circumflex artery originated from the right coronary artery. Subsequently, the ostium of the two coronary arteries was excised from the pulmonary artery wall with "coronary buttons", i.e. an area of arterial tissue. Afterwards, they were sutured with 7.0 Prolene posteriorly in the orifices of the neo-aorta, without undue traction or torsion of the coronary arteries.

The defect in the pulmonary artery following coronary artery excision was repaired with a Cormatrix patch. Then the Lecompte maneuver was performed, through which the pulmonary trunk was repositioned anterior to the aorta.

The great vessels were anastomosed using 7.0 Prolene and reinforced with Bioglu. A simple suture was used to close the atrial septal defect, the heart cavities were refilled, and the

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cross clamp was removed with satisfactory perfusion.

Cardiopulmonary bypass time was 157 minutes while the aortic cross clamp time-ischemia time was 112 minutes.

Given the extent of this cardiac procedure, the myocardial edema and the risk of bleeding, we decided to delay the sternum closure, a maneuver called "open chest", until the hemodynamic, respiratory and metabolic parameters were stabilized. A plastic tube (polyvinyl) was used as a chest spreader, over which heterologous bovine pericardium was sutured and protected by Steri-Drape.

In the first 3 hours postoperatively, due to clotting disorders, approximately 200 ml of blood was drained through the chest tubes having a significant hemodynamic impact, hence the hemostatic intervention was decided in the ICU. Meanwhile, NovoSeven and thrombocytes were administered.

Regarding the functioning of the urinary system, diuresis in the first hours was unsatisfactory, ranging between 2-5ml per hour, with the growth of potassium, which required placement of a peritoneal dialysis catheter.

The first 24-hour status was severely altered, with persistent metabolic acidosis, supraventricular tachycardia with narrow complexes requiring therapeutic and medium to high doses of inotropic medication consisting in combined adrenaline, dopamine, noradrenaline and milrinone.

The following days were hemodynamically steady with normalization of ventilation and metabolic parameters, regain of diuresis. Therefore, at 8 days after surgery on the 22nd of March 2013, the chest was closed.

However, the evolution was unfavourable in terms of the digestive system, with reduced gastric tolerance, absent bowel sounds and abdominal distension. A pediatric surgery consultation was requested on the 28th of March 2013 and median supra and sub-umbilical laparotomy was done revealing a fecaloid generalized peritonitis, perforation of both cecum and colon, with cecum necrosis, and mesenteric perforation. An atypically ileocecal resection and ileostomy was performed, the mesenteric tear was sutured, and peritoneal lavage and drainage was employed.

On 29th of March 2013, the newborn presented a central catheter dysfunction at the level of the right internal jugular vein with clinical and ultrasound suspicion of thrombosis on the right superior vena cava, which is why the surgical preparation of the left saphenous vein was required. Evolution slowly improved, with restored bowel transit on the ileostomy and decreased doses of inotropic drugs.

On April 3rd, 2013, abdominal surgery was reinitiated for dissecting bowel adhesions.

In terms of respiratory system, after physical examination, x-ray and transthoracic echography, a suspicion of a right diaphragmatic paresis arose, and on 16th of April 2013 the right hemidiaphragm was plicated via anterolateral thoracotomy.

After this, a period of steady evolution follows, during which the patient develops several episodes of generalized tonic-clonic seizures.

From the medication point of view, we mention: Meronem, Targocid, Colistin, Metronidazole, Tazocin, Trienam, Vancomycin, Diflucan, Amiodarone, Humaglobin 5%, Dexamethasone, Furosemid, Spironolactone, Controloc, Motilium, Aspatofort, Biotics.

On 09th of May 2013, the pediatric surgical team reevaluated the status and considered it appropriate to take out the ileostomy and perform end-to-side ileo-colic anastomosis and partial great omentum resection.

Regarding the cardiological status, echocardiography showed good contractility, with no valvular dysfunction of the

neo-aorta or neo-pulmonary artery, or pleural or pericardial effusion.

On 21st of May 2013, after 68 days of hospitalization, the patient is discharged with stable cardio-respiratory, metabolic, renal and digestive status.

At 5 year-follow-up and after all the severe complications that occurred during hospitalization, the patient is in a good medical condition without any organic distress or neuro-psychic disorders with good acquisitions and social abilities (figure no.1, no.2.)

Figure no. 1. Postoperative scars at 5 year-follow-up



DISCUSSIONS

The immediate few days following the procedure presented a high cardiovascular and metabolic risk, due to the inflammatory effects of post-extracorporeal circulation, myocardial edema and clotting disorders. Generally, these factors lead to occurrence of post-hemorrhagic cardiac tamponade.

Due to hypotension and inotropic support with vasoconstricting effects, the urinary system progressively limits its function, requiring renal replacement therapy with peritoneal dialysis, mainly preferred in our clinic.

The "open chest" acts like a medical and surgical "compromise" and it is maintained depending on the hemodynamic stability with significant decrease of inotropic support, negative hydric balance (200-300ml) for 24-48 hours and thoracic blood drainage under 75ml/ 24h.(3) The procedure of chest closure is generally performed in the pediatric intensive care unit without influencing the infectious risk.(4)

Visceral complications such as ischemia may occur as a result of extracorporeal circulation effects, by affecting the microcirculation or as a result of inotropic medication with vasoconstrictive effect. Also, the placement of a peritoneal dialysis catheter may have a direct mechanical impact on the intraabdominal visceral segments.(5)

Thrombogenic episodes commonly found in the superior vena cava can be attributed to lesions and sutures at the level of the cannulation site combined with the effect of procoagulant medication used for treating clotting disorders.(6)

Dissection maneuvers in the aortic arch area or prolonged ventilation may cause phrenic nerve injuries leading to diaphragmatic paresis clinically showed by tachypnea, polypnea, or the need for invasive or non-invasive respiratory support.

Non-pulsatile cerebral circulation, hypothermia and neuroleptic medication can cause neurological imbalances or seizures.(7)

CONCLUSIONS

In case of a complex pathology with a generalized

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effect on the cardiac, hepato-renal, pulmonary, metabolic and hematologic system, the ideal medical and surgical therapy is sustained by a multidisciplinary team. However, there is the possibility of complications secondary to malperfusion and multiorgan immaturity.

Early diagnosis, experience and appropriate treatment are the main target of this multidisciplinary team in order to improve their performance with each treated patient.

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