A CASE OF TRUNCUS ARTERIOSUS TYPE II*

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A case of truncus arteriosus type II is reported. Truncus arterious is an uncommon congenital cardiac defect where a single great vessel exits the heart. Truncus arteriosus is usually fatal, if untreated. This defect occurs when the conus arteriosus and the truncus divide erroneously in the embryo. Palliative surgery in truncus arteriosus has been unsuccessful. Pulmonary banding has been tried and was ineffective and usually fatal. We operated on a nine-month-old (6200 g) male infant with a type II (Edwards-Collett) defect and a large ventricular septal defect. The pulmonary artery average pressure was 51 mmHg. We performed a cardiopulmonary bypass in the usual manner. Pulmonary arteries were resected from the truncal root, and primary end-to-end anastomosis of the truncal root to the ascending aorta was performed. Right ventricle to pulmonary artery continuity was provided using a valveless Gore-Tex graft. We lost our patient due to intractable pulmonary hypertension on the first postoperative day. *Key words: truncus arteriosus, cyanotic heart defects, total correction.*

Truncus arteriosus is an uncommon congenital cardiac defect in which a single great vessel stemming from the heart branches into the coronary arteries, pulmonary arteries and the ascending aorta. Truncus arteriosus is a usually fatal congenital defect which, without treatment, results in 65 percent mortality in the first three months of life due to untreatable congestive heart failure[1]. Truncus arteriosus accounts for approximately two to four percent of all congenital cardiac anomalies[2]. The single trunk in truncus arteriosus imposes an appallingly high flow and elevated pulmonary artery pressure which leads to the development of heart failure early in life. Run off from the truncal artery creates a low diastolic pressure causing a decreased coronary perfusion and a further decrease in ventricular performance. The presence of a truncal valve regurgitation further complicates the picture[2].

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This defect occurs in the embryo when the conus arteriosus and the truncus divide erroneously and the spiral septum develops deficiently. Because the spiral septum also takes a role in the closure of the ventricular septum, truncus arteriosus always occurs concomitantly with a large ventricular septal defect (VSD). Works of Garcia-Peláez and colleagues[1] concluded that the truncus arteriosus appears in the chick embryo, at 50-56 hours of incubation. They strongly believe that there is a possibility that the defect occurs in a similar way in mammals, including the human embryos[3].

In some cases of truncus arteriosus the VSD can include the tricuspid annulus and cause difficulties in closure. Unfortunately, efforts to palliate truncus arteriosus in patients with Collett and Edward's Type I or II defects by pulmonary artery banding in an attempt to decrease pulmonary blood flow yielded poor results, with most series reporting mortalities in the range of 50 to 60 percent[4]. The follow-up patients with truncus arteriosus showed a high incidence of early pulmonary vascular disease. Thus, the pioneering article of Ebert and colleagues[4] concluded that physiological correction should be performed early in life.

Case Report

We operated on a nine-month-old (6200 g) male infant who had truncus type II (Edwards-Collett) defect. The patient upon his cardiac catheterization was found to have a large VSD (Fig. 1), truncus arteriosus type I defect which, on perioperative examination, was found to be actually a type II defect. His pulmonary artery average pressure was 51 mmHg. The arcus aortae was normal

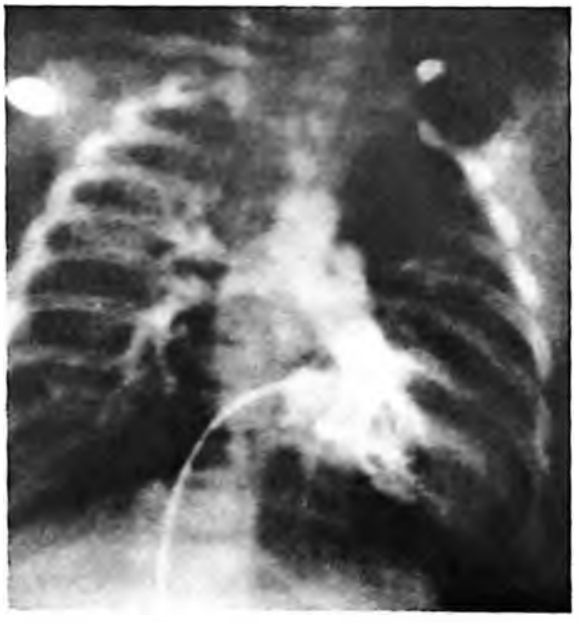

Fig. 1: Angiography showing truncus arteriosus.

and there was no patent ductus arteriosus (PDA). The patient had been operated on previously at five months of age, when a pulmonary artery banding had been performed to the left pulmonary artery.

Operational technique

After standard median sternotomy exposure, a cardiopulmonary bypass was conducted in the usual manner. We used systemic hypothermia. The ascending aorta was cannulated above the bifurcation of the truncus, and both vena cava were cannulated.

The left and right pulmonary arteries were exiting the aorta via different roots from the posterior part. We first performed a debanding of the left pulmonary artery, and then performed an aortotomy (Fig. 2). The pulmonary arteries were resected from the truncal root in continuity such that sufficient tissue surrounding the orifices of the pulmonary arteries was present for distal conduit anastomosis. Some truncal tissue had to be removed, leaving a defect at the posterior part of the future aorta. This was later repaired using a Gore-Tex (polytetrafluoroethylene) patch. Truncal valve of the ventricular outflow tract was bicuspid. There was no truncal regurgitation.

We followed guidelines of Ebert and colleagues[4], who reported that when making an initial incision in the posterior wall of the aorta, caution is necessary to avoid the suspension system of the truncal valve, and that the orifice of the left coronary artery should be identified. The commissures of the truncal valve are usually

Fig. 2: Intraoperative view of repair of truncus arteriosus. VSD is seen through right ventriculotomy, and truncus is divided.

short and the depth of the Valsalva's sinus is less than normal. Thus, it is possible that the left coronary orifice is high in the sinus and can be incised at the time of exploration of the pulmonary cuff, or that it could be incorporated into the suture at the time of closure of the aortotomy[4].

A primary end-to-end anastomosis of the truncal root to the ascending aorta was performed. Closure was carried out using a running polypropylene suture. Care was given to protect the coronary ostia. In our case we had no coronary anomalies. Afterwards, we performed a longitudinal incision to the right ventricle exposing the VSD. It was taken far enough to expose the VSD so that the ventricular opening would be adequate for the conduit. The defect was presenting as a subaortic defect, which is usual. The defect was closed in such a manner as to leave the aorta on the left side of the heart. We performed a right atriotomy such that no atrial septal defect or patent foramen ovale was present. The tricuspid valve was normal. We later used a number 14 Gore-Tex (PTFE) tubular graft to create the right ventricle outflow tract (Fig. 3). After weaning from cardiopulmonary bypass, the patient's arterial pressure was 80 mmHg. We used a full dose of dopamine (15 µg/kg/min) for support. Despite intensive therapy to prevent hypertensive crisis, we lost our patient on the first postoperative day due to the intractably high pulmonary hypertension.

Discussion

The techniques for repair of truncus arteriosus are individualized according to the specific valve morphology[5]. If there is truncal valve regurgitation, which is frequent in patients with truncus arteriosus, perioperative and late mortality are increased.

Most authors suggest that repair should be undertaken before 100 days of life when incidence of pulmonary hypertension and pulmonary hypertensive crises in the postoperative period is low[6,7]. In our case, the resistance was high with the patient at nine months of age.

Following the contributions of Ebert and colleagues[4], several reports have shown improvement in the outcome of patients undergoing operations in which complete repair and physiological correction are carried out. Survival in the first month of life is low, and thus many authors, including Sharma et al.[8], Di Donato et al.[9], and Ziemer et al.[10], continue to recommend postponing the operation until the patient is two to three months of age. According to Bove et al.[2], it is significant to mention that medical treatment is useless and further decreases the chance of the infants to gain weight, nor does it protect them from infections or pulmonary vascular disease or left ventricular dysfunction. Bove et al.[2] also point out that a valve in the pulmonary position provides a substantial hemodynamic improvement. Thus, beginning in 1986, valved conduits have been used.

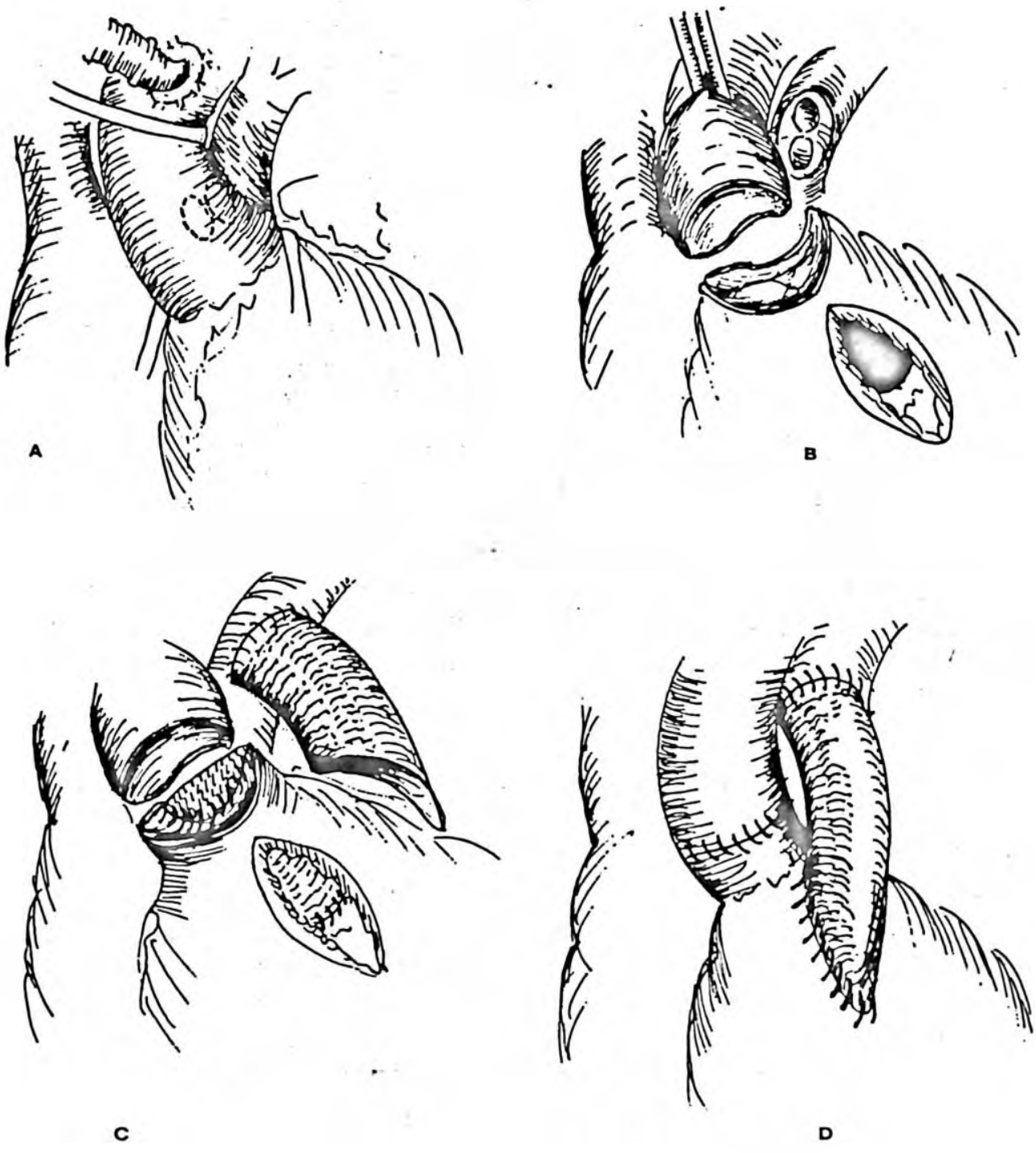

Figure 3: procedure of repairing type II truncus arteriosus.
A : preoperative schematic view of type II truncus arteriosus. Pulmonary arteries origin is seen in the posterior aspect of the truncus.
B : The VSD is seen through the right ventriculotomy. After the completely transecting of the truncus, the pulmonary arteries origin is dissected free and is separated from the aorta.
C : The VSD is closed with a patch. The distal anastomosis of the conduit to the origin of the pulmonary arteries and restoration of the posterior wall of the truncus by a patch is seen.
D : Completed repair. The aorta end - to end anastomosis is seen. The anastomosis from the right ventriculotomy to the origin of the pulmonary arteries by a conduit is seen.

Fig 3

There are four important preoperative risk factors which increase mortality in patients with truncus arteriosus[6]: 1-truncal valve regurgitation, 2-associated interrupted aortic arch, 3- coronary artery anomalies, and 4-repair undertaken after 100 days of life.

We had one (4th) of the four risk factors which many authors describe as the most vulnerable.

Repair of truncus arteriosus is most favorably undertaken within the first days of life[6], when incidence of pulmonary vascular hypertension is low. According to Ebert et al.[4], a valved conduit is preferable in the initial operation. They claim that since there are great swings in pulmonary artery pressure due to unstable resistances in the pulmonary vascular bed of an infant, a rapid increase in pulmonary artery pressure in a non-valved conduit will have an increased amount of pulmonary regurgitation and can cause sudden right ventricular failure. On the other hand, if the size of the pulmonary artery is near normal or normal for age, then a non-valved conduit seems rational[4].

Petz et al.[1] suggest that a conduit valve is not essential in the correction of truncus arteriosus even in the neonate. But they also say that a functional pulmonary valve is necessary if the pulmonary vascular resistance is high.

Repair using a valveless conduit for the right ventricular outflow tract is recommendable for infants without high pulmonary vascular resistance and who do not have right ventricular failure. Valveless conduits may cause pulmonary regurgitation, increasing the pulmonary resistance, and resulting in insufficient right ventricle output and immature pulmonary vasculature[11]. In our case a previous pulmonary banding had been performed to prevent this.

Delaying surgery to such time when a valveless conduit is suitable and the pulmonary vascular resistance is low is risky, as was seen in the case of our patient. Because this causes a hyperactive pulmonary vasculature before surgery, postoperative mortality is high. Patent foramen ovale should be retained in the neonatal reconstructions to aid right ventricular output.

Barbero-Marcial et al.[12] proposed an alternative technique for type I and II truncus cases, which should be noted. They used a corrective operative technique without use of an extracardiac conduit, and directly anastomosed the pulmonary arteries and the right ventricles, the anterior wall being constructed with a patch with a monocuspid valve.

Emphasis should be given to keeping the pulmonary vascular resistance minimized. Acute pulmonary artery hypertensive crises are difficult problems to encounter. Maintenance of high oxygen saturation and respiratory alkalosis, continuous sedation and paralysis, and avoidance of unnecessary tracheal suctioning help minimize pulmonary resistance.

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