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Truncus Arteriosus with Anomalous Origin and Intramural Course of the Right Coronary Arterie: A Surgical Challenge

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

The *truncus arteriosus communis* is a very complex congenital cardiac malformation occurring in less than 1% of all congenital heart disease patients. Early repair remains one of the most challenging procedures in congenital cardiac surgery. Due to improvement in surgical reconstruction strategies over the past years, there is an acceptable short and long-term survival for those patients. However, each new case represents a "*high mountain to climb*" in ambition to achieve physiological repair with acceptable hemodynamic and a good long-term survival. We therefore present a very special case with anomalous origin and intramural course of the right coronary artery.

Keywords: Truncus arteriosus; coronary artery; surgery; cardiac malformation.

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1. INTRODUCTION

The *truncus arteriosus communis* is a complex congenital cardiac malformation occurring in less than 1% of all congenital heart disease [1,2]. Anatomically, a great single vessel arises from both ventricles supplying the aorta, the pulmonary arteries and the coronary arteries. The *truncus arteriosus* is always associated with a large sub truncal ventricular- septal defect. Further associated pathologies include atrial septal defect (90%), anomalous origin of the coronary arteries (40%-50%), interrupted aortic arch (25%) and persistent left superior vena cava (5-10%) [3,4]. However, the incidence of patients with intramural coronary arteries is very low (0.1%-0.3%) [5]. There are two major classifications used to describe the anatomy, those of Collet and Edwards [6] and those of van Praagh [2]. McGoon and colleagues performed the first surgical repair with the use of a valved conduit in 1967. Due to improvement in surgical reconstruction strategies over the past years, there is an acceptable short and long-term survival for those patients. However, each new case represents a surgical challenge. Due to the presence of an intramural coronary artery, the risk of acute myocardial ischemia and sudden death increases dramatically perioperatively. We therefore present a very special case with anomalous origin and course of the coronary arteries and discuss our surgical strategy.

2. CASE PRESENTATION

A 34-day old boy with diagnosis of *truncus arteriosus* and large subtruncal ventricular septal defect was admitted to our centre for surgical repair. The pre- operative course was uneventful without any hemodynamic support. The preoperative echocardiogram at our institution confirmed the diagnosis. There was no evidence of severe coronary anomalies. Therefore, we performed no CT scan and no cardiac catheterization for evaluation of the coronary arteries. The complex repair was performed from a midline sternotomy with aorto- bicaval cannulation and cardio- pulmonary bypass at systemic temperature of 24°C. After dissection of the pericardium, we detected the anomalous right coronary artery. The vessel arises very high anterior above the valvular plan and took an intramural course. (Fig. 1). The left coronary artery originated dorsal between the left and non-coronary sinus and took course also intramural. The pulmonary arteries were snared at the onset of bypass, and cold St. Thomas cardioplegia (4°C, 35 ml/kg) was delivered through the truncal vessel after placement of the cross- clamp. The heart arrested immediately and the coronaries were filled sufficient with cardioplegia solution. The pulmonary arteries were excised from the *truncus arteriosus* and exposed through a very high horizontal anterior incision. We had to do the dissection very carefully to prevent the coronaries from severe injury. We checked the right intramural coronary artery with a small probe and detected no severe stenosis and no injury. Before right ventricular outflow tract reconstruction, we performed an end- to end-patch- anastomosis from the dilated truncus arteriosus to the very small aortic arch. For the next step, the ventricular septal defect was approached through a longitudinal ventriculotomy and closed with a glutaraldehyde pre-treated pericardial patch. A bovine jugular venous valve conduit (Contegra® 12 mm) was used for right ventricular outflow tract reconstruction.

With moderate inotropic support the patient was weaned from cardio pulmonary bypass without signs of myocardial ischemia and with acceptable pulmonary pressure. He was administered in a stable hemodynamic constitution to our intermediate care unit. The postoperative course was uneventful and the patient was discharged to the ward with a lower level of care five days later.

3. DISCUSSION

Neonatal *truncus arteriosus* repair remains one of the most challenging procedures in congenital cardiac surgery. The first successful repair was performed in 1967. Due to innovative techniques und surgical strategies in some specialized centres, the early mortality ranges between 5%-10% [7]. Concomitant ventricular septal defects and anomalies of the coronary arteries could complicate surgical course and therefore determine the postoperative outcome dramatically [8]. Tlaskal and colleagues determined coronary artery anomalies as one of the main risk factors for death [8]. Great care must be taken to ensure that there is no deformation of the coronary ostia between truncal vessel and the aortic arch. In case of coronary distortion and tension, myocardial ischemia would lead to severe biventricular failure with unsuccessful weaning from cardio-pulmonary- bypass [9,10]. Therefore, recognition of potential coronary artery anomalies is essential and must be considered in every new case [11].

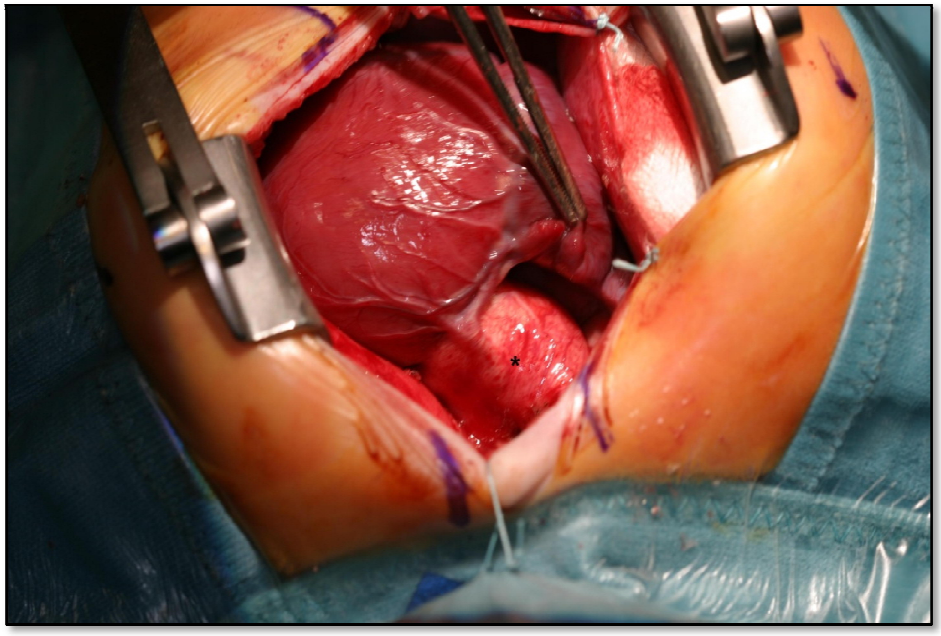

Fig. 1. The right coronary ostium arises anterior and very high from the truncus wall*

4. CONCLUSION

In conclusion, our experience shows that early *truncus arteriosus* repair is feasible with excellent results. However, complex cases with concomitant coronary artery anomalies must repair with the highest level of care and experience.

CONSENT

Written informed consent was obtained from the patient for the publication of this paper and any accompanying images.

ETHICAL APPROVAL

As the report is a case presentation formal ethics approval is not applicable. The report has been conducted in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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