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REVIEW

Transposition of the great arteries: Rationale for tailored preoperative management

Transposition des gros vaisseaux : rationnel pour une prise en charge préopératoire sur-mesure

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Summary As preoperative morbi-mortality remains significant, care of newborns with transposition of the great arteries is still challenging. In this review of the literature, we discuss the different treatments that could improve the patient's condition into the preoperative period. Instead of a standardized management, we advocate personalized care of these neonates. Considering the deleterious effects of hypoxia, special attention is given to the use of non-invasive technologies to assess oxygenation of the tissues. As a prolonged preoperative time with low cerebral oxygenation is associated with cerebral injuries, distinguishing neonates who should undergo early surgery from those who could wait longer is crucial and requires full expertise in the management of neonatal congenital heart disease. Finally, to treat these newborns as soon as possible, we support a planned delivery policy for foetuses with transposition of the great arteries.

Abbreviations: ASO, arterial switch operation; BAS, balloon atrial septostomy; CHD, congenital heart disease; IVS, intact ventricular septum; NIRS, near-infrared spectroscopy; PGE1, prostaglandin E1; PPHN, persistent pulmonary hypertension of the newborn; PVL, periventricular leukomalacia; rSO_2, regional oxygen saturation; TGA, transposition of the great arteries; VSD, ventricular septal defect.

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MOTS CLÉS
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Spectroscopie dans le proche infra-rouge ;
Switch artériel

Résumé La morbi-mortalité préopératoire des nouveau-nés ayant une transposition des gros vaisseaux est relativement conséquente, ce qui fait que leur prise en charge reste difficile. Dans cette revue de la littérature, nous discutons des différentes thérapeutiques qui peuvent améliorer l'état préopératoire de ces patients. Plutôt qu'un traitement standard, nous prônons une prise en charge personnalisée de ces nouveau-nés. Du fait des effets néfastes de l'hypoxie, une attention toute particulière est portée à l'utilisation de techniques non invasives de monitorage de l'oxygénation tissulaire. Puisque la durée de l'hypoxie cérébrale préopératoire est prédictive de l'intensité des lésions cérébrales, la distinction des nouveau-nés qui doivent pouvoir bénéficier d'une chirurgie précoce de ceux qui peuvent attendre plus longtemps est cruciale. Ceci requiert un niveau d'expertise important dans le domaine des cardiopathies congénitales. Enfin, nous prônons une politique d'accouchement programmée en cas de transposition des gros vaisseaux afin de traiter ces nouveau-nés le plus rapidement possible.

Introduction

Transposition of the great arteries (TGA) is the most common cyanotic congenital heart defect (CHD) presenting in the neonatal period, accounting for 5—9% of cardiac malformations [1,2]. In TGA, the ventriculo-arterial connection is discordant, which means that the aorta arises from the morphological right ventricle, and the pulmonary artery arises from the morphological left ventricle (Fig. 1). The pulmonary and systemic circulations are therefore in parallel rather than in series. As the deoxygenated blood is recirculated through the body (right ventricle—aorta connection) — whereas the oxygenated blood recirculates through the lungs (left ventricle—pulmonary artery connection) — at least two of the three possible communications between the pulmonary and systemic circulations are obligatory to support early survival: a patent ductus arteriosus, an atrial septal defect (always present during pregnancy) or a ventricular septal defect (VSD) (optional). Thus, TGA can be categorized based on the presence or absence of VSD. Usually, newborns with TGA with an intact ventricular septum (IVS) become cyanotic in the first days of life when the ductus arteriosus closes; among these, patients with reduced mixing opportunities (TGA-IVS with restrictive foramen ovale and/or closure of the ductus arteriosus) become symptomatic with extreme cyanosis early after birth. Leading inevitably to progressive hypoxia and acidosis, TGA is an almost always fatal when left untreated.

The physiological and anatomical correction of TGA is the finest example of the successful evolution of the surgical treatment of CHD. Indeed, the advent of the arterial switch operation (ASO) allowed better postoperative survival and outcomes than atrial switch procedures [3]. However, and even if data are scarce, preoperative mortality (describing a fatal adverse evolution during the time between birth and surgery) of newborns with TGA ranges from 3.6% to 10.3% [4—6]. In comparison, in a retrospective study involving 19 European institutions, operative mortality was reported to be 6% [7]. Since the widespread use of balloon atrial septostomy (BAS) and prostaglandin E1 (PGE1) therapy, no new major technique — with the exception of extracorporeal circulatory assistance, which is fortunately rarely used — has improved the postnatal condition of these newborns.

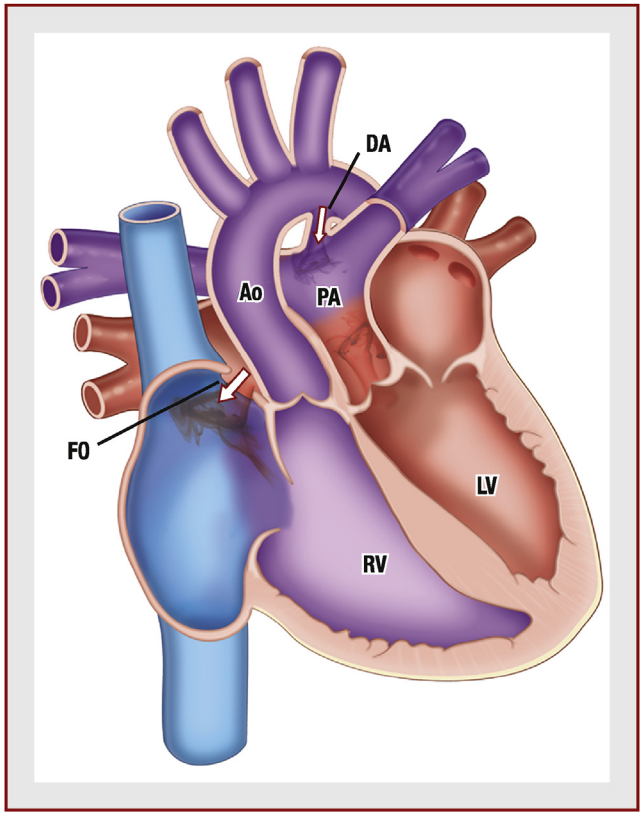

Figure 1. Schematic representation of transposition of the great arteries with an intact ventricular septum at birth. The aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle. Thus, aortic blood saturation is poor and newborns become cyanotic. At birth, two foetal communications exist concomitantly: the foramen ovale and the ductus arteriosus. Ao: aorta; DA: ductus arteriosus; FO: foramen ovale; LV: left ventricle; PA: pulmonary artery; RV: right ventricle.

Nevertheless, current non-invasive technologies allow us to closely monitor these patients and to identify those who will probably benefit from early surgery. Conversely, ASO can be delayed in certain circumstances. In this review of the literature, we discuss the different aspects of the preoperative management of newborns with TGA.

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