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REVIEW

Management of patients with pulmonary atresia, ventricular septal defect, hypoplastic pulmonary arteries and major aorto-pulmonary collaterals: Focus on the strategy of rehabilitation of the native pulmonary arteries

Prise en charge des atrésies pulmonaires avec communication inter-ventriculaire, hypoplasie des artères pulmonaires et collatérales : mise au point sur la stratégie de réhabilitation des artères pulmonaires

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Rehabilitation

Summary Pulmonary atresia with ventricular septal defect (VSD), hypoplastic native pulmonary arteries (PAs) and major aorto-pulmonary collateral arteries (MAPCAs) is a rare and complex congenital cardiac disease. In broad outline, two surgical approaches are available for patients with this condition. The first is characterized by one or several stages of complete unifocalization of the supplying MAPCAs, with or without incorporation of the native pulmonary arteries (PAs), connection of the right ventricle to the 'neo-Pas' and, if possible, concomitant or delayed closure of the VSD. The second strategy is based on rehabilitation of the native pulmonary arteries. The first step is a direct right ventricle to native PA connection, to promote the growth of native PAs. The establishment of antegrade flow also allows an easier approach for

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interventional catheterization, enabling dilatation or stenting of the stenosis and then closure of the communicant collaterals. When the development of the native PAs is satisfactory, the complete repair is performed. If it is necessary to suture a MAPCA to the PA ('unifocalization'), this is accomplished by connecting the collateral artery to an already developed native branch. Our team developed this multidisciplinary strategy with good results. Based on this experience as well as on the published literature, we describe this strategy of management of patients with pulmonary atresia, VSD, hypoplastic pulmonary arteries and major aorto-pulmonary collaterals (MAPCAs).
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MOTS CLÉS
Atrésie pulmonaire ;
Collatérales ;
Réhabilitation

Résumé L'atrésie pulmonaire avec communication inter-ventriculairev (CIV), hypoplasie des artères pulmonaires natives et collatérales est une cardiopathie congénitale rare et complexe. Il existe schématiquement deux approches chirurgicales différentes. L'une est représentée par une chirurgie d'unifocalisation des collatérales, incorporant ou non les artères pulmonaires natives, la connexion du ventricule droit aux « néo » artères pulmonaires et si possible dans le même temps, la fermeture de la CIV. La seconde stratégie est basée sur la réhabilitation des artères pulmonaires natives. La première étape consiste en une ouverture de la voie ventricule droit — artère pulmonaire, dans le but de promouvoir la croissance des artères pulmonaires natives. L'établissement d'un flux antérograde rend ainsi les APs accessibles aux procédures de cathétérisme interventionnel. Lorsque le développement des APs natives est satisfaisant, la réparation complète peut être réalisées, avec unifocalisation des collatérales et fermeture de la CIV. Notre équipe développe cette stratégie multi-disciplinaire avec de bons résultats. À partir de cette expérience et de celle retrouvée dans la littérature, nous proposons une description et une mise au point de la stratégie de réhabilitation des artères pulmonaires dans la prise en charge des patients avec atrésie pulmonaire, hypoplasie des artères pulmonaires, CIV et collatérales.
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Background

Pulmonary atresia with ventricular septal defect (VSD), hypoplastic native pulmonary arteries (PAs) and major aorto-pulmonary collateral arteries (MAPCAs) is a rare and complex congenital cardiac disease. In broad outline, there are two different surgical approaches to these patients. One of these approaches is characterized by one or several stages of complete unifocalization of the supplying MAPCAs, with or without incorporation of the native PAs, connection of the right ventricle (RV) to those 'neo-PAs' and, if possible, concomitant or delayed closure of the VSD [1,2].

The other approach is based upon rehabilitation of the native PAs [3]. The first step is to create a direct connection from the RV to the native PAs, in order to promote native PA growth. Establishment of antegrade flow allows also an easier approach for interventional catheterization, making dilatation/stenting of stenosis and then closure of communicant collaterals possible. When the development of the native PAs is satisfactory, the complete repair is performed. If it is necessary to suture a MAPCA to the PA ('unifocalization'), this is accomplished by connecting the collateral artery to an already developed native branch.

Our team developed this multidisciplinary strategy with good results [4]. Based on this experience as well as on the literature, we review this strategy for the management of patients with pulmonary atresia, VSD, hypoplastic PAs and MAPCAs.

Which patients are we talking about?

Anatomical description

To enter this category, a patient should present with pulmonary atresia, no persistent arterial duct and 'hypoplastic' but present central PAs. This excludes patients with 'normal' PAs vascularized through a ductal structure, with the atresia being simply at the valvular level or more complex. It excludes, of course, patients without any central PAs. In real life, it is not always easy at first examination to differentiate clearly between a tortuous arterial duct and an MAPCA but it is clear that none of these patients should be prostaglandin E1-dependent. 'Hypoplastic' PAs have to be determined according to the Nakata index [3] (below 90 mm/m^2 [or 100 mm/m^2, depending on the authors]). None of these patients should be readily accessible to complete repair without unifocalization, which means that a normal output will not be able to flow through the central PA before rehabilitation.

The origin of the pulmonary flow is completely due to the MAPCAs. The distribution of the native PAs and the MAPCAs to the pulmonary segments has to be appreciated and evaluated.

The location of the VSD is the same as in Tetralogy of Fallot — a conoventricular position. The anatomical investigations have to confirm that it is a single VSD. Furthermore, the position of the coronary arteries has to be checked. The aortic arch may be left- or right-sided, which may have importance for the surgical plans.

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