Standardization of reporting would help to define best treatment for pulmonary atresia, ventricular septal defect and major aortopulmonary collateral arteries

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Keywords: Congenital heart disease • Follow-up study • Pulmonary atresia • VSD • MAPCA

It is no surprise that most clinicians still have doubts on the best procedures to offer to their patients with pulmonary atresia, ventricular septal defect (VSD) and major aortopulmonary collateral arteries (MAPCAs). The morphological features and clinical presentation of those affected vary greatly as the condition represents a spectrum of disease. The condition is rare and most teams have customized their care to individual patients. Under the brand name of ‘unifocalization strategies’ are gathered various operations and none of the previous reports of large series of unifocalization allows the delineation of outcomes of specific procedures [1–3]. These series describe their overall philosophy of management rather than the results of specific procedures in specific patient settings. In this view, the report in this issue of the experience of the Fuwai Hospital is enlightening [4].

First of all, their recruitment is such that they are able to gather an experience at a much faster pace than other units, apart from maybe the teams from Stanford and Birmingham, who have specialized recruitment for this condition. We are still in a field where practice makes excellence, especially for rare conditions, and it is likely that their team will quickly become expert in its management. As in other fields, they have adopted a strategy that had the best chances to be reproducible, reliable and efficient. They proceeded with the rehabilitation of small central pulmonary arteries with a right ventricle to pulmonary artery conduit, which gave them access for repetitive interventional catheterization procedures to the central pulmonary arteries. The principle of this approach has already been described but they added original contributions [5, 6]. They proceeded with the aggressive occlusion of all MAPCAs, which presented as a dual blood supply to lung segments at the same time of the initial procedures, at the time of the repair or in the interim period to decrease competitive flow in these territories and facilitate growth of the pulmonary arteries. Most of these procedures were performed as hybrid procedures in the catheterization laboratory. The occlusion of these MAPCAs did not seem to be related to adverse events such as desaturation.

Additionally, all procedures between the first conduit and the repair were made by interventional catheterization. The results of this well-described uniform strategy were satisfactory. The mortality rate was minimal: Two patients died of pneumonia, one of myocarditis and one of right heart failure. Apart from this latter mortality directly related to the strategy (and that is likely acceptable in the beginning of such a strategy), the survival of this difficult subset of patients was excellent. The post-repair ratio between the right and left ventricular pressures was similar to the one described by most teams. At this stage, their rate of repair may seem slightly inferior to some others, but they clearly described that they are currently performing repair in the restricted population with the most favourable pulmonary anatomy. They could clearly enlarge their indication for repair, but that may be at the cost of some mortality. Alternatively, they might increase their rate of repair by performing additional procedures. In yet unpublished results of our strategy of rehabilitation of the central pulmonary arteries with repetitive surgeries, our rate of repair was 73% [7].

The results of a strategy have to be interpreted in the light of the population operated. Some of the patients of Fuwai hospital seem to have undergone their rehabilitation strategy at an older age than usually seen and it is possible that some of these patients may be selected survivors of the initial neonatal period. They rightly opted to exclude patients with absent central pulmonary arteries. These patients represent a mixed group of patients. Some have well-developed MAPCAs and intrapulmonary pulmonary vessels and present with heart failure and high saturation at birth and usually have favourable outcomes without unifocalization. Others have multiple small tortuous MAPCAs as the sole pulmonary blood supply and have much worse outcomes.

In this group of patients, they have however integrated some with no MAPCAs individualized and some with patent ductus arteriosus. There is no strict consensus on the definition of this condition, but most have tended to restrict their series to patients whose pulmonary blood flow exclusively depended on collaterals,
eliminating those with patent ductus. In my experience, patients with pulmonary atresia and ductus-dependent lung circulation have larger pulmonary arteries and more favourable outcome. This did not seem to be the case in the Fuwai experience but remains difficult to ascertain. This team has the enormous merit of having adopted a clearly delineated uniform strategy in a well-defined subset of patients. This publication undoubtedly increases our knowledge; however, as practitioners, we would be able to more accurately interpret the currently published experiences if all had used a standardized way of reporting. We need each series to describe the morphological features of the patients in the same way, to specify whether MAPCAs are counted as trunks or separate branches and to delineate the exact procedures performed and the nature of the materials used. Until we achieve better standardization of the reporting of our experiences, our progress in the identification of the best strategies for this condition will be slow.

**Funding**

Yves d’Udekem is an NHMRC Clinician Practitioner Fellow (1082186). The Victorian Government’s Operational Infrastructure Support Program supported this research project.

**Conflict of interest:** Yves d’Udekem is a consultant for companies MSD and Actelion.

**REFERENCES**


