The Fuwai hospital experience with patients presenting late with pulmonary atresia, ventricular septal defect and hypoplastic pulmonary arteries

Yves d’Udekem

The reported experience from the team of Fuwai Hospital with rehabilitation procedures of the pulmonary arteries performed beyond 6 months of age in patients born with pulmonary atresia, ventricular septal defect (VSD) and major aortopulmonary collateral arteries (MAPCAs) shed new light on the potential of these techniques to achieve repair when these patients are diagnosed later in life [1]. Their experience should be followed closely because their access to a very large population is likely to make them acquire expertise in this field more rapidly than anywhere else: in a period of only 3 years they treated close to 40 of these older patients, more than any other center in the world could hope to see.

As they adequately emphasized, patients presenting after 6 months of age are a selected group of patients promised to a better survival than their younger counterparts, a fact that we also noticed in our previous experience. The authors demonstrated that, as patients with this condition grow older, an increasing proportion of collateral arteries become stenotic, likely causing the early death of some of the patients at the worse end of the spectrum of the disease. Some of the patients studied presented signs of severity of the disease: close to a third of them had no central pulmonary arteries. Others seemed to have a more favourable condition with half presenting with a persistent ductus, a characteristic usually absent in patients with lung circulation exclusively dependent on collateral flow. The quality of their results is undoubtedly remarkable. In a relatively short period, up to 80% of the patients could hope to undergo final repair with no mortality directly related to the procedures or the condition. This undoubtedly constitutes an argument demonstrating that rehabilitation procedures of the native pulmonary arteries are effective in this age group.

The authors adopted an aggressive approach towards the occlusion of the collaterals arteries before or at the same time as the surgeries. All MAPCAs not bearing stenosis were occluded if they brought dual blood supply and if their occlusion did not cause significant desaturation. This aggressive policy has some benefits. It avoids the competition of flow between the collateral arteries and the forward flow in the native vessels. We have noticed in our past experience that this flow competition can impede the growth of the native vessels. By decreasing the pulmonary artery pressures, it also undoubtedly helped to reduce right ventricular pressures after completion of the repair. We have so far been reluctant to adopt such a systematic policy in Melbourne in order to avoid any compromise to their pulmonary blood supply.

Surgeons are divided between partisans of unifocalization and those who support rehabilitation of the native pulmonary vessels [2, 3]. The present work demonstrates clearly the feasibility of rehabilitation procedures in patients presenting late with pulmonary atresia, VSD and hypoplastic pulmonary arteries. There remain, however, several unsolved questions in the management of these difficult patients and they are all revolving around the fate of collateral arteries once integrated in the recreated pulmonary circulation. Under the generic term of ‘unifocalization’ the teams who have promoted the mobilization of all collateral and native vessels in a recreated pulmonary circulation have used procedures aiming at the rehabilitation of the native pulmonary vessels and translocation of collateral arteries, but the contribution of both have never been specified. Until we obtain a careful longitudinal study of the fate of the operated collateral arteries in unifocalization procedures in a similar way that native pulmonary vessels were observed in this manuscript, we will still be uncertain of the final contribution of these vessels to the pulmonary circulation. We have, in Melbourne, performed unifocalization of collateral vessels in a very restricted number of cases where these vessels seemed to be of the same nature and size as pulmonary vessels as they enter the lungs. But we still reject final repair for some patients and leave them to palliation when their pulmonary circulation is limited to only small collateral arteries and all our attempts to rehabilitate a native circulation have failed. Until we understand the fate of trans-located collateral arteries, we will still remain uncertain whether there is a subset of patients who can never be repaired, and we will not know how to identify these patients and how early they could potentially be identified.
Conflict of interest: Yves d’Udekem is a Career Development Fellow of The National Heart Foundation of Australia Research Program (CR 10M 5339). The Victorian Government’s Operational Infrastructure Support Program supported this research project.

REFERENCES

