Editorial Commentary

Mechanical mitral valve replacement in children: an update

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Provenance: This is an invited article commissioned by the Section Editor, Xicheng Deng (Department of Cardiothoracic Surgery, Hunan Children's Hospital, Changsha, China).

Comment on: Ibezim C, Sarvestani AL, Knight JH, *et al.* Outcomes of Mechanical Mitral Valve Replacement in Children. Ann Thorac Surg 2019;107:143-50.

Submitted Aug 11, 2019. Accepted for publication Aug 27, 2019.

doi: 10.21037/tp.2019.08.03

View this article at: http://dx.doi.org/10.21037/tp.2019.08.03

Ibezim *et al.* have recently brought serious methodology to assess mitral valve replacement (MVR) in children (1). MVR has been a matter of discussion for a long time. It is always a heart-breaking decision to make, especially in young children.

Mitral valvuloplasty is the preferred method of managing infants and children with congenital malformations of the mitral valve. Unfortunately, when mitral valvuloplasty fails, a mechanical MVR (MMVR) is performed because there is no other sustainable option. To date, mechanical prostheses have indeed a low profile, smaller size and enhanced durability compared to xenograft valve, which are associated with very bad short-term results in children.

When facing this situation, it is crucial to rely on data to help with family counselling and team decision-making. Outcomes concerning MVR and MMVR in children is often limited to small size samples or single-institution experiences, although reference articles are to be acknowledged (2-4).

Ibezim and colleagues from the Pediatric Cardiac Care Consortium (multi-institutional study) reviewed 441 patients, younger than age 21 years, undergoing mechanical MVR, with 144 patients age <2 years. Median age was 4.3 years and early mortality was 11.1%. To our knowledge, there is no such paper with this number of patients and a 17 years follow-up. They must be congratulated for this massive work of data collection and data processing. They have delivered intelligible messages which will be useful in everyday practice.

Creating 4 groups of age (<2; 2–6; 6–12; 12–21 years) reveals the higher risk of immediate mortality in young patients, under 2 years of age. Other risk factors were identified: CAVC defects, Down syndrome, high size/weight ratio. These results confirm what had been already shown in other studies: historically, placement of a mechanical mitral valves, in patients younger than age 2 years, has been associated with high mortality rates. But, once postoperative period is passed, long-term mortality of young patients (<2 years) was also associated with lower, long-term, death/transplant-free survival. Contrary to other previous study, with reported excellent long-term results for patients who survive the hospital period (2,5), long-term mortality was high, with 76% survival at 20 years and with 88 deaths out of 392 patients (22.5%) in this series. This points out the global severity of mitral issues in children.

In consequence, the mortality results are a little disappointing, not only in the short-term but also and notably in the long-term period. Also, we would like to make sure that the readers understand the long-term outcomes used in this study. The outcome, "mortality due to CHD" or heart transplant, are not complications related to the MMVR itself. It is more related to the severity of the initial pathology leading to high-risk risk surgery and long-term cardiac attrition.

It is to be emphasized that every single patient is a different story, especially when dealing with congenital heart diseases. When it comes to MVR, the situations are very different between a L-TGA and an unbalanced AV canal in a Down patient. Addressing data concerning one single surgery, in very different patients, carries pitfalls of misunderstanding or underestimating some phenomenons.

Thus, Ibezim *et al.* analysed CAVC, PAVC, L-TGA and Shone Syndrome separately. They have identified a higher risk of short-term mortality and long-term attrition for CAVC defects, especially when associated with Down syndrome.

Also, analyzing effects of MVR on survival, within different diagnoses, was very interesting. MVR does not affect long-term outcomes for patients with L-TGA and PAVC receiving MMVR, compared to patients with L-TGA and PAVC who did not require MMVR; but death/transplant-free survival was lower after MMVR in patients with CAVC. This indicates that the prognosis is more influenced by the pathology itself, than the surgery of MVR. It is to be known when counseling families.

After reading this very interesting article, some questions remain unanswered:

(I) First question unanswered: diseases of circulatory system are responsible for long-term mortality, in 21 patients out of 77. We assume that it is mainly due to cerebral complications of long-term anticoagulation (stroke, cerebral haemorrhage). The difficulty of achieving optimal anticoagulation in children, and especially in the youngest, may lead to complications such as valve thrombosis, hemorrhage or strokes. To realize the long-term consequences of MMVR, searching for non-lethal cerebral complications would have been informative and could guide medical teams when informing families before performing MMVR.

(II) Second question unanswered: is MMVR sometimes performed too late? At the time of MVR, patient's condition is usually precarious, especially in infants, with very high pressure in the left atrium and permanent need of oxygen, despite optimal medical treatment. Sometimes, patients can't even be weaned from ventilator. MMVR is then a measure of last resort: patients have been operated upon several times and myocardium has been enduring unfavorable load conditions for a long period of time; ventricular function can be severely impaired, after several surgeries, with severe mitral dysfunction in-between. Obviously, it has a negative impact on short- and long-term results. And it is an understatement to say that mechanical MVR are performed in bad general conditions. Unfortunately, and of particular importance, was the unavailability of data such as the severity of symptoms on which the decisions to proceed with mechanical MVR were made; and in our opinion, it would have been interesting to know how many surgeries had been performed before MMVR, especially in infants or children requiring valve surgery during the first 2 years of life.

We support Ibezim *et al.* when writing that mechanical MVR was commonly performed because of availability of smaller sizes prosthesis and lack of durable alternatives. We want to insist on the lack of alternatives, which has not dramatically changed for the last 20 years. First of all, mitral repair techniques have improved. Some authors suggest that mitral repair is almost always feasible in expert centers (6). Other techniques have been developed, such as mitral-aortic annular enlargement, surgical placement of a Contegra (Medtronic Inc, Minneapolis, MN, USA) conduit positioned in a graft tube or Ross II procedure in larger patients: these procedures are exceptionally used in children <2 years, on a case-by-case basis, and large studies will probably never be available.

In infants and children (<2 years old), there is frequently a discrepancy between the prosthetic valve size and the mitral annulus size; the smallest available prosthetic valve is often too large for the native mitral annulus. MVR procedure is then challenging, and is associated with tremendous complications, including complete heart block, LVOT obstruction, risk of prosthetic leaflet entrapment, pulmonary vein obstruction and compression of the circumflex coronary artery. Insertion of a surgically placed modified stented jugular vein graft valve (Melody valve, Medtronic Inc., Minneapolis, MN, USA) was recently reported, and this procedure appears to be a reproducible technique providing satisfactory short-term results (7,8). To date, this technique is probably the best alternative to MMVR in children with the smallest annulus, despite left ventricular outflow tract obstruction related to the length of the Melody valve conduit remains an unknown issue. Recently, the first results of the smallest mechanical valve available suggest that the newly approved 15-mm valve offers a reliable strategy for palliating severe mitral disease in young children. Reasonable hope is therefore allowed to improve outcomes in the youngest children (9).

In conclusion, Ibezim and colleagues have to be congratulated for this study, gathering a huge amount of data and delivering clear messages. In this study, results are consistent with published data concerning short- and long-term outcomes. Furthermore, this confirms, with recent data, ideas that have been known for long (5,10,11). We highlight the fact that issues and outcomes concerning

MVR in young children have remained unchanged for the last 20 years. In consequence, this study, besides helping medical teams in taking decisions, is pointing the fact that emerging alternatives and new armamentarium (6,9) have to be assessed in the future, in order to improve outcomes in children, especially <2 years, suffering from mitral diseases.

Acknowledgments

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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Cite this article as: Metras A, Seguela PE, Roubertie F. Mechanical mitral valve replacement in children: an update. Transl Pediatr 2019;8(5):455-457. doi: 10.21037/tp.2019.08.03