

Survival Improvement in Tetralogy of Fallot With Absent Pulmonary Valve: A Nouvelle Cuisine Message



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The absent pulmonary valve is a rare form of Tetralogy of Fallot (ToF) in which the pulmonary valve is lacking and severe pulmonary valve regurgitation is present from fetal life. The severe regurgitation of the pulmonary valve determines a wide range of pulmonary artery dilatation that in the most severe cases causes compression and obstruction of the tracheobronchial tree since the neonatal period. Surgical repair is needed to address all ToF features but also reducing pulmonary artery dilatation with the adjunct of pulmonary artery plication. Survival has been generally reported to be poor compared to the other ToF forms, especially for those patients in which perioperative ventilatory support and/or neonatal repair is required.^{1,2} In this particular subset of patients, a significant cause of increased mortality is represented by tracheobronchomalacia that is responsible for severe respiratory distress and difficulties in weaning ventilatory support.

In their study, Dorobantu et al reported³ outcomes of 98 UK patients with ToF with absent pulmonary valve who underwent repair between 2000 and 2013. The study is elegantly executed and reported and is probably the largest cohort of such patients in the recent era. The population was heterogeneous in terms of age and presentation at surgery; 15% were neonates and 13% required preoperative ventilation. Follow-up information regarding reoperation and status was obtained from the National Institute for Cardiovascular Outcomes Research (NICOR), which retains information on each patient operated on in the United Kingdom. One of the most important findings was a reduction of late mortality (8%) compared to previous studies (20%).^{1,2,4} Despite the improvement in survival, reoperation rates for the pulmonary valve or for the pulmonary artery remain a significant burden in the long-term affecting around 40% of the patients. Neonatal repair and



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Central Message

Tetralogy of Fallot with absent pulmonary valve showed a better survival after surgical repair. Airway involvement is a risk factor for mortality indicating an area to address in future improvements.

preoperative mechanical ventilatory support were identified as risk factors for mortality as already reported, reiterating the key role of airways structure in the survival of this subset of ToF patients. The paper should be read by all surgeons and cardiologists and the evidence provided should be used when counseling parents regarding long-term outcome in the current era. On the other hand, the paper reminds us of a nouvelle cuisine dish: beautiful presentation, perfect execution, and an explosion of flavors. Your eyes are completely satisfied by only looking at it, your mouth is astonished when tasting it but the stomach will never be happy and will ask for more. Nouvelle cuisine resembles the food perfection in a small size portion, gives you immediate palate happiness but you still want more. The same happens with the Dorobantu work: you will remain satisfied by the reading, the analysis, and the reassuring results of this elegant study but the stomach of a clinician will ask for more information like: (a) how was it possible to reduce mortality?, (b) are the results better because patients were selected (meaning that inoperable patients were excluded by the nature of the study)?, (c) how were the airways?, and (d) which surgical techniques or right ventricle to pulmonary artery prosthesis were used? It was not possible to find any of this information in the paper due to the nature of the dataset from which it was

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retrieved, as acknowledged by the same authors. The lack of these important pieces of data weakens the overall importance of the paper because the improvement of the survival reported cannot be explained with clinical data and we remain unable to learn how it was possible in the United Kingdom to achieve such results. In conclusion, Dorobantu et al demonstrate important evidence of improved results for a rare variation of ToF. It should be used as a starting point for more in-depth research on how was achieved such improvement, so may be we will be able to expand widely their results and improve patients care.

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