See Article page 365.

Commentary: Ross procedure is indeed a safe and durable option for critical aortic valve disease in neonates and infants

Osami Honjo, MD, PhD

Management of neonates and infants with critical aortic valve stenosis (AS) poses a significant challenge. The patients present with various degree of left ventricular dysfunction and endocardial fibroelastosis due to critical downstream obstruction, secondary pulmonary hypertension, and important additional lesions such as mitral valve dysplasia and aortic arch obstruction. Balloon aortic valvuloplasty has been a standard first intervention for these patients, but surgical intervention is needed when balloon valvuloplasty is ineffective or results in significant aortic insufficiency (AI). Ross/Ross-Konno procedure is an effective surgical treatment for those neonates and infants with residual critical AS or developed significant AI, but is associated with relatively high mortality and morbidities, especially among patients who have aortic arch obstruction and/or mitral valve disease, severe left ventricular dysfunction, or require an emergency or urgent Ross procedure due to hemodynamic instability.¹

Laxford and colleagues² from The Children's Hospital at Westmead report their institutional experiences of Ross/Ross-Konno procedure spanning more than 2 decades. They report no early mortality and only 1 late mortality among 35 neonates ($n = 13$) and infants ($n = 22$). This is a shockingly excellent outcome for patients who have so many high-risk anatomic and physiological features. The first hurdle is to execute

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CENTRAL MESSAGE

Luxford and colleagues report excellent early and late outcomes of Ross/Ross-Konno procedure in neonates and infants, including preserved neo-aortic valve function and geometry.

Ross/Ross-Konno procedure without any major residual anatomic lesions. The majority of the patients in their series had trivial or mild neo-AI. Only 1 patient required concomitant mitral valve repair—a low prevalence compared with other clinical series that may be partly associated with this excellent survival because we all know significant mitral valve disease in this population is an Achilles' heel. The second hurdle is to effectively support a dysfunctional left ventricle after Ross/Ross-Konno procedures. The combination of preexisting left ventricular dysfunction and/or endocardial fibroelastosis and prolonged cardiopulmonary bypass time poses a scenario of significant postoperative left ventricular dysfunction. Six (17%) patients required postoperative mechanical circulatory support, but all of them were weaned from support at a mean duration of 4.7 days. In our institution, we sometimes employ a hybrid procedure (bilateral pulmonary artery banding and ductal stenting) as an interim procedure for patients with critical AS and severely reduced left ventricular function. Lastly, the authors report no reintervention to the neo-aortic valve during the follow-up period and 1 patient required surgical reintervention for left ventricular outflow tract. The neo-aortic valve remains competent in the vast majority of the patients and neo-aortic valve z score remains constant at the final follow-up, which is consistent with other series.³

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This excellent clinical report is very encouraging for all of us who treat this critically ill patient cohort. The report emphasizes that early technical success leads to excellent early and late outcomes. I am particularly encouraged by the fact that all patients who needed mechanical support postoperatively were able to be weaned. Only 1 subgroup is missing from this report: The group with significant mitral valve disease, who remain a high-risk group with less-optimal options and outcomes.

See Article page 365.

Commentary: Ross procedure in neonates and infants: Withstanding the litmus test of time

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More than 50 years after Donald Nixon Ross developed his eponymous procedure of replacing a diseased aortic valve with pulmonary autograft, the technique has become an established surgical option for aortic valve replacement.¹ The Ross procedure offers superior hemodynamic parameters in comparison to alternative means for aortic valve replacement, avoidance of anticoagulation therapy, and growth potential for patients at younger ages. In the pediatric population, it is the sole option if valve repair is not feasible or fails. The main disadvantages of the Ross procedure are autograft failure primarily in the form of neo-aortic root dilatation and an almost definite need for reintervention on both valves down the line.

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Manan Desai, MD, Michael Ma, MD, and Can Yerebakan, MD

CENTRAL MESSAGE

Ross procedure in infants and neonates entails high initial morbidity. However, the long-term outcomes are good, with an acceptable, but definite risk of reintervention(s).

Luxford and colleagues² have shared their excellent results of the Ross/Ross-Konno procedure in patients younger than age 1 year. We congratulate the authors for their outstanding results that should encourage us to utilize the technique more often in pediatric populations. In other series, the average early mortality of the Ross procedure in infants varies from 7% to 27%.³⁻⁶ In this retrospective analysis of 35 patients, there was no surgical mortality. Therefore, we have the privilege to comment on a study with among the best early surgical results for Ross/Ross-Konno in infants and neonates. Strikingly, 60% of all patients underwent emergency procedures; all of the 13 neonates had an emergency repair. Morbidity was high: use of extracorporeal membrane oxygenation (17%), delayed sternal closure (70%), and incidence of low cardiac output (60%). There are other notable aspects of this study. Compared with previous studies, fewer patients