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Editorial

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Atrial Septal Defect with Down Syndrome and Postsurgical Pulmonary Hypertension

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▸ See the article "Reversibility of Pulmonary Hypertension Following Surgical Atrial Septal Defect Closure in Children with Down Syndrome" in volume 27 on page 247.

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Conflict of Interest
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Down syndrome (DS) which is the most common chromosomal anomaly, occurs in 1/ 319–1,000 live births.[1] Half of them have congenital heart disease, atrioventricular septal defect (AVSD, 45%), ventricular septal defect (VSD, 35%), secundum atrial septal defect (ASD, 8%), and tetralogy of Fallot (TOF, 4%).[2]

In Korea, the prevalence of DS was 4.4 per 10,000 total births (1.5%). Congenital heart defect (CHD) was identified in 57% in DS patients and ASD was the most common (30.5%), followed by VSD (19.3%), patent duct arteriosus (17.5%), and AVSD (9.4%).[3]

DS is thought to be strongly associated with pulmonary hypertension (PH) in patients with CHD, in whom PH develops more rapidly. Besides pulmonary vascular structural changes, many respiratory problems are seen in DS patients, including large tongue, small hypopharynx, tonsils and adenoid hypertrophy, laryngomalacia in addition to gastroesophageal reflux, immunologic dysfunction, and pulmonary hypoplasia known to cause airway disease.[4] An imbalance of mediators like nitric oxide, thromboxane A_2, and prostacyclin also may be seen.[5]

Bush et al.[6] reported 13 autopsy findings from individuals with DS who died of cardiopulmonary diseases, in which alveolar simplification, signs of persistence of a double capillary network in the distal lung, prominent bronchial vessels, and recruitment of intrapulmonary bronchopulmonary anastomoses may worsen gas exchange in patients with DS.

In a recent Japanese nationwide congenital heart surgery database for the repair of CHD with DS, 167 patients had ASD with DS. Mean age at operation was 2.1 ± 4.2 years, and preoperative cardiac catheterization was done in 143 patients (85.6%). Thirty-three patients (23.1%) showed pulmonary vascular resistance > 4 WU/m^2, and 9.8% of patients took PH medication. PH medication at discharge was needed in 21.3% of patients and nitric oxide inhalation was needed in 6.6% undergoing surgery.[7]

The risk and outcomes after open heart surgery were similar in both patients with and without DS.[8]

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In an analysis of the United States national database, DS was not associated with a significant mortality risk, although post-operative morbidity was common in congenital cardiac surgery. Length of stay was prolonged in patients undergoing ASD closure, VSD closure, and TOF repair, which were associated with post-operative respiratory and infectious complications.[9]

Although operating severe PH is controversial, successful percutaneous closure of ASD following therapy with intravenous prostacyclin was reported in a patient with irreversible pulmonary hypertensive arteriopathy.[10]

How about the long-term results of surgical treatment of adult patients with ASD? Horvath et al.[11] reported a late mortality rate of 4.9% and a late morbidity rate of 12.4%, mainly because of arrhythmias in their 166 consecutive patients with a mean follow-up period of 90 months. There were 2 operative deaths due to respiratory failure and severe PH respectively, and 13% of patients had perioperative complications. Patients with systolic pulmonary artery pressure > 30 mmHg showed a higher late mortality rate.

In the current study by Lee et al.[12], 15 children with DS underwent ASD patch closure successfully, which included 3 patients with severe PH. It was possible to discontinue selective pulmonary vasodilators in 2 patients, and one patient was continued on selective pulmonary vasodilators because moderate degree PH continued due to a chronic lung problem.

Postsurgical PH in patients with ASD and DS is sometimes unpredictable or even worse than expected. Nevertheless, with the aid of selective pulmonary vasodilators, their lesions can be repaired successfully.

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