Poster | 染色体異常・遺伝子異常

Poster (I-P01)

Chair:Kazushi Yasuda(Department of Pediatric Cardiology, Aichi Children's Health and Medical Center)
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[I-P01-07]VSDに対する心内修復術を施行した18trisomy患児の予後

○内山 敬達[1], 根本 慎太郎[2], 岸 勘太[3], 吉村 健[4] (1.高槻病院 小児科, 2.大阪医科大学 胸部心臓血管外科, 3.大阪医科大学 小児科, 4.関西医科大学 小児科)

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(Background)Congenital heart defects (CHD), particularly ventricular septal defect (VSD) is highly common in patients with trisomy 18. However,indication for cardiac surgery for trisomy 18 is controversial due to poorly documented post-surgery prognosis. (Patients and Methods)To investigate the prognosis of intra cardiac repair (ICR) in these patients, we evaluated medical data from 30 patients with trisomy 18 in three institutes in Japan. (Results) Sixteen (53 %) of 30 were performed cardiac and palliative surgery with their careful parent' s content. 15 patients were VSD and 1 patient was coarctaion of aorta with VSD. Twelve (75%) of 16 patients with VSD underwent ICR and survived. Eleven (91%) of 12 patients underwent pulmonary artery banding (PAB) between 12 to 126 days (median, 72 days) before ICR while one patient underwent primary ICR. The remaining four (25%) of 16 patients underwent PAB, where only one patient survived. ICR was performed between 150 and 570 days after PAB (median, 321 days). Post ICR survival ranged from 450 to 2,550 days (median, 1095). Nine (75%) of 12 patients who underwent ICR also underwent tracheostomy after PAB. These 12 patients did not have gastrointestinal anomaly such as esophageal atresia. (Conclusion)In this study, 53 % of patients with trisomy 18 underwent cardiac surgeries and all of whom underwent ICR for VSD survived. Hence, ICR for VSD may help to prevent cardiac deaths which could lead to higher survival rate in trisomy 18 with congenital heart defect.