Substantial Growth of Atretic Pulmonary Artery after Repair of Total Anomalous Pulmonary Venous Connection combined and Congenital Diaphragmatic Hernia

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Abstract

Total anomalous pulmonary venous connection (TAPVC) with congenital diaphragmatic hernia (CDH) is a disease entity with high mortality rate. Association with atretic left pulmonary artery increased the complexity of the anomalies. Here, we reported a newborn baby with these complex congenital anomalies successfully treated surgically. Over 13 years after surgery, there was substantial growth of left pulmonary artery which was angiographically atretic at his newborn stage, which was rarely reported. Currently, this patient is drug free and is in functional class I of New York heart association.

Introduction

Total anomalous pulmonary venous connection (TAPVC) with congenital diaphragmatic hernia (CDH) is a disease entity with high mortality rate[1], for which only 13% of the patients could survive to discharge.[1] Severe respiratory compromise will happen after birth. Association with atretic left pulmonary artery increased the complexity of the anomalies. Here, we reported a newborn baby with these complex congenital anomalies successfully treated surgically and growth of atretic pulmonary artery was shown after long-term follow-up.

Case Report

A male newborn, birth body weight 2.2kg, suffered from respiratory distress with desaturation and tachycardia after birth at other hospital. Endotracheal tube was inserted on the next day after birth and then he was transferred to our hospital. With ventilator FiO2 50%, his SpO2 could be kept around 90%-95%. Chest x ray film showed haziness of left lung field where bowel gas was noted (Figure 1A). Congenital diaphragmatic hernia (CDH) was diagnosed. Polysyndactly of left thumb was also noted. After general condition was stabilized, repair of left hemi-diaphragm was performed 2 days after birth. During surgery, a left postero-lateral defect of diaphragm about 5cm x 3cm was noted. The intra-abdominal organs were pulled down. The defect was repaired with prolene suture continuously. No patch was used. The extra-thumb was also excised. However, after surgery for diaphragm repair, his SpO2 was around 91%-93% with ventilator FiO2 70%. Chest x ray showed whiteout of left lung field. Besides, systolic cardiac murmur was also found. Echocardiography was checked on the next day after CDH surgery (3 days after birth), by which cardiac type of TAPVC, atrial septal defect (diameter 0.54cm), patent ductus arteriosus (PDA) and hypoplastic left pulmonary artery (LPA diameter 0.21cm, RPA diameter 0.34cm) and impaired left ventricular function (LVEF 46%) were disclosed. Cardiac catheterization was done 4 days after birth and showed severe pulmonary

hypertension (LV pressure 58/2mmHg, MPA pressure 45/15mmHg) and angiographically atretic diminutive LPA (Figure 1B & 1C). Cardiac repair was performed 6 days after birth. After median sternotomy, cardiopulmonary bypass (CPB) was performed with aortic and bi-caval cannulation. PDA ligation was done at first. After incision of right atrium, a large atrial septal defect (diameter 1cm) and an enlarged coronary sinus ostium were seen. Coronary sinus was un-roofed, which produced a large opening between coronary sinus and left atrium. A patch with fresh autologous pericardium was used to close the ASD and CS, by which pulmonary and cardiac venous flow were directed to the left atrium. All the procedures were performed under CPB without circulatory arrest. On the other hand, because the LPA was grossly atretic, there were no anything could be done for that. Sternum was approximated immediately after surgery. Temporary epicardial pacemaker had been used for 4 days after cardiac surgery because of transient atrioventricular block. This patient recovered uneventfully. Endotracheal tube was removed 7 days after cardiac surgery. He was discharged from the hospital 21 days after cardiac surgery. Echocardiography was arranged 5 months after cardiac repair and showed normal PV return, no residual ASD and normal LVEF (57.8%). When he was 1 year old, he received another surgery for pyloric stenosis and adhesion ileus.

During follow-up, left lung was still hypoplastic (Figure 2A). CT scan was followed-

up 13 years after repair of CDH and TAPVC and showed substantial growth of LPA from angiographically atretic lesion to 4mm in diameter (Figure 2B). Currently, he is pinkish, drug free and in functional class I of New York heart association.

Discussion

CDH is a disease entity with high mortality rate which was close to 70% at 1 year.[2] Regarding associated lesions, 17% of CDH infants had congenital heart disease (CHD) and the association was stronger (27~28%) if still-births and pregnancy termination were included.[1] Among the cardiac lesions associated with CDH, total anomalous pulmonary venous connection (TAPVC) was relatively rare (1.8%).[1] For the infants with CDH associated with TAPVC, the mortality rate was further higher and only 13% of the patients could survive to discharge.[1] For our patient, cardiac anomaly was diagnosed after CDH repair. According to The CDH EURO Consortium Consensus – 2015 Update[3], the grade of recommendation is D to perform echocardiography within the first 24 hours after birth. In fact, echocardiography is very useful and helpful for the infants with CDH because the incidence of associated cardiac anomalies is not low. In addition to TAPVC, angiographically atretic LPA was present in our patient when he was born. However, substantial growth of the angiographically atretic LPA was shown by computed tomography 13 years after surgery for anomalies of diaphragm and heart, which seems rarely reported.

Conclusion

TAPVC with CDH, left lung hypoplasia and atretic left pulmonary artery is a rare disease entity with a very critical condition. After successful surgical repair, it could not be sure whether atretic pulmonary artery has the potential of growth until long-tern follow-up is done.

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Figure 1. (A) Chest x ray film of our newborn patient showed bowel gas in left chest cavity; (B) no left pulmonary artery was shown by antero-posterior view of angiography of pulmonary artery; (C) no left pulmonary artery was shown by lateral view of angiography of pulmonary artery

Figure 2. (A) Chest x ray film 13 years after surgery; (B) Substantial growth of angiographically atretic left pulmonary artery to 4mm in diameter by computed tomography