Total Anomalous Pulmonary Venous connection combined with Congenital Diaphragmatic Hernia and Left Lung Hypoplasia

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Introduction:
Total anomalous pulmonary venous connection (TAPVC) with congenital diaphragmatic hernia (CDH) and left lung hypoplasia is a rare disease entity. Severe respiratory compromise will happen after birth. Here, we reported a newborn case with these complex problems successfully treated surgically. Currently, over 13 years after surgery, the patient is a teenager now and is in functional class I of New York heart association.

Method:
A male newborn, body weight 2.2kg, suffered from respiratory distress with desaturation after birth. Endotracheal tube was inserted and the ventilation was kept acceptably by the effort of pediatric doctors. Chest x ray showed haziness of left lung field where bowel gas was noted. CDH was diagnosed. After general condition was stabilized, repair of left hemi-diaphragm was performed at 4 days of age. During surgery, a left posterolateral defect of diaphragm about 5cm times 3cm was noted. The abdominal organ was pull down. The defect was repaired with prolene suture continuously. However, after surgery, the patient was still desaturated. We arranged echocardiography which showed cardiac type of TAPVC, atrial septal defect, patent ductus arteriosus (PDA) and hypoplastic left pulmonary artery (LPA). We arrange cardiac repair for him at 7 days of age. TAPVC repair with PDA ligation were performed smoothly. Because the LPA was very diminutive, we could not do anything for that. Sternum was approximated immediately after surgery. This patient recovered uneventfully. When he was 1 year old, he received another surgery for pyloric stenosis and adhesion ileus.

Result:
Currently, he is over 13 years old with good activity and is in functional class I of New York heart association and is pinkish. CT scan was followed up 13 years after repair of CDH and TAPVC and showed growth of LPA from diminutive size to 4mm in diameter.

Conclusion:
TAPVC with CDH and left lung hypoplasia is a rare disease entity with a very critical condition. Surgery is the only treatment of choice. If the patient can stand the surgeries well, the long-term survival can be anticipated.