PW4-5

Topsy-Turvy Heart: Very rare rotational heart anomaly with tracheobronchial anomalies

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Introduction: Topsy-Turvy heart is characterized by approximately 90 degree clockwise rotation of the entire heart as a block around the axis from apex to base, leaving the right ventricle in a superior spatial relationship to the left venticle, while the great arteries are displaced inferiorly and posteriorly into the distal mediastinum resulting elongation of the brachiocephalic arteries. As a result of the rotation trachea and bronchi become stretched and elongated. To date, only 4 alive cases have been described in the literature. We report 3 cases with morphologic details, clinical presentation, additional anomalies and their management.

Methods : The patients were 1 month, 5 months and 4 years of age. Two girls and a boy. Last two patients were siblings born to consanguineous parents. In addition to the characteristic rotational anomaly of the heart and the great vessels, all patients had large aorto-pulmonary window defects, some degree of left main bronchus stenosis and systemic pulmonary hypertension. First two patients with severe heart failure symptoms, underwent open heart surgery. Aorto-pulmonary window defects were closed by using pericardial patch under total circulatory arrest (18°C) in case 1 and under selective antegrade serebral perfusion (26°C) in case 2.

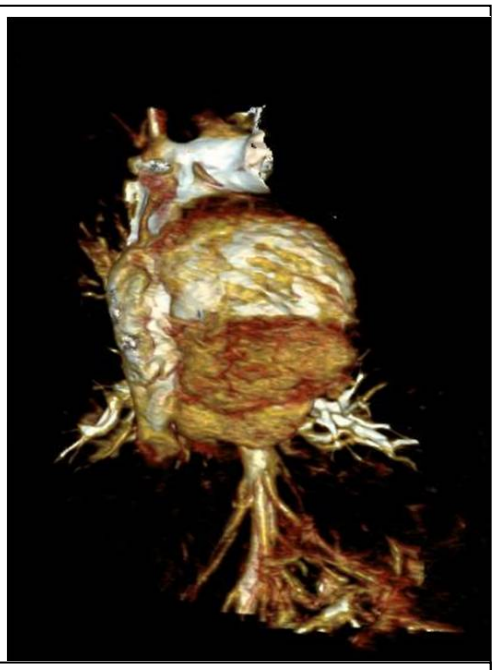

Figure 1:CT angiogram of case1
3D reconstruction, anterior view

Results: Severe respiratory failure and air trapping in the left lung developed in the first case. Sternum was left opened. ECMO support was initiated in postoperative day 10 due to resistant respiratory failure. She died of multiorgan failure in postoperative day 16. The second case had a prolonged intensive care unit stay due to pulmonary hypertensive crisis and postoperative sepsis. He was discharged from the hospital on the 30th postoperative day in good clinical condition. Cardiac catheterization of the third patient revealed high pulmonary vascular resistance (12 Wood U). Bosentan treatment was started and second hemodynamic evaluation was scheduled. Genetic counseling is pending.

Conclusion: Our findings support the previous reports in the literature. Patients have no intracardiac anomaly except a large aorto-pulmonary communication. History of consanguinity , like other report, support a single gene disorder with a recessive mode of inheritance. Respiratory complications which may be fatal in postoperative period, necessitate multidisciplinary team approach.