A miracle pregnancy with unilateral pulmonary agenesis and corrected tracheo-esophageal fistula

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ABSTRACT

Introduction: Congenital unilateral lung agenesis with tracheoesophageal fistula is rare. Even with corrected fistulae, a pregnancy with a single-functioning lung is at risk of respiratory failure, lung infection, pulmonary hypertension, anaesthetic risk, preterm labour and a small baby, leading to a challenging maternity care. **Case Description:** We are sharing a case of a 29-year-old primigravida with congenital left lung agenesis and surgically corrected tracheo-esophageal fistula. Left hemithorax was occupied by hyperinflated right lung, stomach and spleen with elevated left diaphragm. She is kyphoscoliosis, limping, is underweight, yet independent. She presented at 11 weeks of gestation without pre-pregnancy care consultation and baseline investigations. Lung function test at the first trimester was non-conclusive. Echocardiography was normal despite cardiac malposition. No pulmonary hypertension. MDT discussion consisted of O&G Specialist, Maternal Fetal Medicine Specialist, Chest Physician, ORL Surgeons and Family Medicine Specialist, and her wish to continue her pregnancy was supported. Her antenatal care was planned for her and she was advised to inform if any symptoms of respiratory failure. 3 out of 5 pre-delivery ward admissions were due to pneumonia. Despite a normal mid-trimester scan, the fetus developed early fetal growth restriction. She required portable oxygen supplementation due to type-1 respiratory failure from 30 weeks onwards. Elective caesarean section was scheduled at 33 weeks due to worsening respiratory function. She was discharged after 4 days and given a subdermal contraceptive implant. The child was discharged 7 days later. Surveillance is maintained under pre-pregnancy care services. **Discussion:** This is a challenging clinical case that demonstrates an increased cardiopulmonary physiological demand as the pregnancy progresses. Albeit limited scientific evidence is available, MDT care and patients' insight remain a critical component in determining optimal pregnancy outcome even in a rare and high-risk case.