Case Report

Pregnancy management and outcome in a woman with congenital unilateral lung agenesis: A case report

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ABSTRACT

Background: Congenital unilateral lung agenesis is a rare condition in which there is complete absence of one lung, in addition to the associated bronchus and pulmonary vasculature. It can be an isolated finding or present in conjunction with cardiac, gastrointestinal, genitourinary and skeletal malformations. Due to the increasing physiological demands on the body during gestation and parturition, this condition poses significant risks in individuals with this condition. Case: A 26-year-old G1P0 with known right lung agenesis was managed with a multidisciplinary approach by obstetrics, maternal–foetal medicine, pulmonology and cardiology specialists. The patient subsequently had an uncomplicated pregnancy and vaginal delivery. Conclusion: Multidisciplinary care led to an optimal pregnancy outcome in a woman with congenital unilateral lung agenesis. Women with this condition with normal baseline function have a good prognosis in pregnancy.

KEY WORDS: High-Risk Pregnancy, lung agenesis, obstetric

INTRODUCTION

Congenital unilateral lung agenesis is a rare condition that is characterised by the complete absence of a lung along with the bronchus, pulmonary artery and veins. It is often associated with other malformations such as cardiac, gastrointestinal, genitourinary and skeletal. The incidence of right or left lung agenesis is similar. Absence of the right lung is associated with an increased risk of bronchopulmonary and other malformations resulting in higher rate of morbidities and mortality.[1] There is an increased risk of infection with right lung agenesis, likely due to impaired bronchial drainage from the exaggerated rotation of the mediastinum and heart. There is a 50% mortality rate at birth or before 5 years of age, with some patients surviving for more than 40 years.[2] Pregnancy poses significant risks to patients with unilateral pulmonary agenesis due to the physiologic changes that occur during gestation and childbirth. Limited cases of pregnancy in patients with unilateral pulmonary agenesis exist in the current literature.

CASE PRESENTATION

A 26-year-old G1P0 was referred to Maternal Foetal Medicine (MFM) clinic at 20 weeks, secondary to...
spontaneous singleton pregnancy. Her medical history was significant for congenital right lung agenesis with dextrocardia and left lung hyperinflation. Other medical history included pre-gravid obesity and mild intermittent asthma. She was asymptomatic and able to tolerate normal daily physical activity.

Preconceptionally, she underwent cardiopulmonary testing, which revealed normal baseline functioning. A transthoracic echocardiogram (TTE) prior to conception showed dextrocardia and shift of the heart to the right upper area of the chest with a right-sided inferior vena cava (IVC) with mildly reduced left ventricular systolic function and an ejection fraction of 45%–50%. Preconceptional chest X-ray demonstrated the heart’s position in the right side of the chest, opacification of the right hemithorax and relative hypoplasia of the right ribcage. There was no focal parenchymal consolidation identified in the left lung, which partially extended into the right hemithorax [Figure 1]. Computed tomography (CT) scan of the chest revealed thoracic findings consistent with the X-ray findings [Figure 2]. She was also recommended to undergo cardiopulmonary exercise stress testing and pulmonary function testing (PFT), which she did not complete before pregnancy.

All the prenatal labs were completed and noted to be normal. A detailed obstetric ultrasound for foetal anatomy assessment and foetal echocardiogram were completed and noted to be normal. Given her normal baseline lung function, an uncomplicated pregnancy course was anticipated. Recommendations included PFT to assess her forced expiratory volume in 1 s (FEV1) and follow-up with cardiology and pulmonology every trimester. Foetal growth assessment every 4 weeks after 24 weeks and third trimester antenatal foetal surveillance were recommended.

The patient remained compliant with her care with all the specialists. Foetal status remained reassuring with appropriate growth for gestational age with normal amniotic fluid volume. The patient’s cardiac and pulmonary status remained stable throughout the gestation. A multidisciplinary conference (MDC) was coordinated in early third trimester for delivery planning among the MFM, anaesthesia, paediatric cardiology, pulmonology, obstetrics and critical care specialist teams.

Labour induction was electively scheduled at 39 weeks of gestation for a planned delivery. Maternal status was monitored closely with telemetry during the course of labour. Early epidural was administered to avoid the detrimental effects of intrapartum pain. She had an uncomplicated vaginal delivery. Careful monitoring of the patient’s volume status was performed during the intra- and postpartum periods, secondary to concern of volume overload from continuous intravenous fluid infusions and autotransfusion following delivery. Her immediate postpartum course was uncomplicated, and she was discharged home in a stable condition on the second postpartum day. The patient was doing well with no issues at her 6-week postpartum visit. She recovered well from her pregnancy and delivery and was then followed for routine gynaecological care.

**DISCUSSION**

Several maternal physiological changes occur during pregnancy to accommodate the demands of the growing uterus, placenta and foetus. Cardiac changes include an increase in cardiac output of 30%–50% due to peripheral vasodilation, an increase in stroke volume due to increased myocardial contractility, an increase in resting heart rate and a decrease in pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR). The 28% reduction in colloid osmotic pressure to pulmonary capillary wedge pressure gradient, increased cardiac preload and increased permeability of pulmonary capillaries increase the risk of pulmonary oedema. Labour causes further increases in cardiac output, especially during uterine contractions due to the movement of blood into the systemic circulation, and after delivery due to relief of the pressure on the IVC as...
well. Women with compromised or reduced cardiovascular function are, thus, at highest risk of developing pulmonary oedema during the second stage of labour and during the immediate postpartum period.\textsuperscript{[3,4]}

The changes in the respiratory system are especially relevant for a patient with unilateral lung agenesis. The increased level of oestrogen causes the nasopharynx mucosa to become hyperemic and oedematous, leading to chronic nasal congestion. Changes in the shape of the thorax are thought to be related to the relaxed ligamentous structures between the sternum and ribs. The diaphragm rises 4 cm into the thoracic cavity. The respiratory muscle function and inspiratory and expiratory pressures are not affected by pregnancy. The minute ventilation increases, which results in a 50%–70% increase in alveolar ventilation. The effects of progesterone and the central nervous system (CNS) chemoreceptors cause a state of hyperventilation, which leads to an increase in PaO\textsubscript{2} and a decrease in partial pressure of carbon dioxide (PaCO\textsubscript{2}) in the maternal blood, allowing for a gradient conducive to CO\textsubscript{2} transfer from foetal to maternal circulation. The chronic respiratory alkalosis state leads to partial renal compensation by increasing bicarbonate excretion. In the late third trimester, there is a slight decrease in partial pressure of oxygen, which is further decreased in the supine position. There is an increased oxygen requirement during pregnancy up to 40%, as demanded by the growing foetus, placenta and maternal organs. Oxygen consumption during a uterine contraction may triple. Maternal oxygen reserve is decreased due to this increased consumption and decreased functional residual capacity. Apnoea, either caused in sleep or with intubation, can lead to a very rapid decrease in PaO\textsubscript{2}, increase in PaCO\textsubscript{2} and respiratory acidosis. Our patient’s stable cardiorespiratory status prior to pregnancy allowed her body to adapt to pregnancy appropriately without sequelae that would have otherwise occurred in a patient with lower pulmonary reserve.\textsuperscript{[5,6]}

These changes are necessary to support the metabolic and oxygen demands of the foetus; however, the increase in stress on a gravid body is often the source of significant morbidity and mortality in women with compromised systems. For these reasons, a multidisciplinary care approach was of utmost importance to coordinate care among the MFM, pulmonology, cardiology, anaesthesia, and primary obstetrics teams. Limitations of this approach include challenges of incorporating all relevant healthcare providers in the MDC, difficulty in scheduling regular patient follow-ups and the inability of the patient to easily access health care due to financial burden, transportation problems, patient fatigue and lack of education on the necessity of frequent visits. However, due to the success of managing this patient’s pregnancy and delivery, we advocate the utility of the MDC and frequent follow-up and testing with each specialty in ensuring the best possible chances for a successful pregnancy outcome in a patient with such a unique and rare condition.

Data availability
Data from this case report cannot be accessed due to confidentiality of patient information. Relevant imaging, laboratory and clinical findings are described above.

Ethics
The case report is IRB exempted. Patient consent was obtained prior to the writing of the case report.

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Conflicts of interest
There are no conflicts of interest.

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