A medical team at Osaka University Hospital, Japan, has conducted successful treatment for the fetal lung disorder Congenital Pulmonary Airway Malformation (CPAM), also known as Congenital Cystic Adenomatoid Malformation (CCAM). By performing a series of prenatal and postnatal treatments, the infant was discharged from the...
hospital without subsequent complications. This series of treatments potentially provides another option for families with unborn babies with the same disorder.

Recent years have seen an increase in diagnoses of CPAM, a fetal lung disorder that can be diagnosed via ultrasound examination. While small lesions have a minor risk of becoming problematic during the fetal stage, severe cases may result in the development of fetal hydrops, a severe edema caused by circulatory insufficiencies in the fetus, thereby raising the risk of fetal death. In such cases, prenatal treatments such as a thoracoamniotic shunt (placement of a tube between the amniotic space that surrounds the fetus and the expanded fetal lung cyst to reduce cyst size) and steroid administration to the mother are conducted. After birth, necessary treatment may involve intensive medical care and operations due to respiratory failure caused by incomplete lung development. Such a series of treatments and operations requires not only the application of advanced techniques but also the necessary facilities, limiting the number of medical institutions where these can be conducted. The medical team at the Center for Fetal Diagnosis and Treatment, Osaka University Hospital (Center Director: Prof. Hiroomi Okuyama), is equipped with the necessary facilities and expertise, and now reports the successful treatment of a severe case of CPAM.

In this case, the fetus showed a large lesion and the additional complication of fetal hydrops. Since there was a risk of fetal death, prenatal treatment was conducted. First, the fetal cyst was punctured, and at the gestational age of 23 weeks, a thoracoamniotic shunt was placed, leading to a decreased cyst volume. However, exacerbation recurred as delivery approached, so the baby was born in serious condition. After birth, the newborn was placed under artificial respiration in the intensive care unit. However, a continually worsening lesion caused the respiratory condition to deteriorate, so the baby was placed under extracorporeal membrane oxygenation (ECMO) one day
after birth. ECMO provides respiratory and circulation support in cases of respiratory and cardiovascular failure by placing a catheter to the blood vessel and circulating part of the blood via the catheter to an external apparatus in which the blood is being enriched with oxygen and pumped back into the body. At two days of age, the newborn underwent a partial lung resection of the lesion of the left lower lobe of the lung while remaining connected to the ECMO. This removed the compression on the healthy lung and the heart, leading to respiratory and circulatory improvement, so that at four days old, the newborn could be removed from the ECMO apparatus. At 14 days, the residual lung lesion was removed (left lower lobectomy), and at 29 days, artificial respiration was no longer necessary. On May 21st, the baby (58 days old) was discharged from the hospital without any subsequent complications.
The above case is the first report of a successful series of prenatal and postnatal treatments for a severe case of CPAM in Japan. This was possible because the Center for Fetal Diagnosis and Treatment at Osaka University Hospital is equipped with the necessary facilities and advanced medical technology. The Center was set up to provide advanced medical treatment from the fetal stage to newborns and provides safe and appropriate treatment with the aim of leaving no subsequent complications. The reported series of treatments may provide another option for families with unborn babies suffering from the same disorder.
Resection of residual lung lesion (Left lower lobectomy)

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Weaning of mechanical ventilation

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Discharge from the hospital in intact survival

Fig. 3

Provided by Osaka University


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