

## 胎児正中心を呈した胸部異所性腎合併先天性右横隔膜ヘルニアの出生前診断例

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## 抄 録

胸部異所性腎は稀な先天異常でしばしば先天性横隔膜ヘルニア (Congenital Diaphragmatic hernia: CDH) を合併するが、胎児診断例は少ない。胎児正中心を呈し右胸部異所性腎、右 CDH の診断に至った症例を経験した。症例は 32 歳、1 妊 0 産。妊娠 18 週の健診時に正中心を指摘した。28 週の超音波検査で右胸腔内構造が左肺より低輝度を示し腫瘍性病変を疑った。MRI 検査で胎児右胸腔内に右腎と肝右葉の陥入を認め、右 CDH と診断した。36 週の肺胸郭断面積比 0.24、肺断面積児頭周囲長比 2.47 と肺低形成の程度は強くないことが予測されたが、CDH に準じて管理を行う方針とした。妊娠 37 週に帝王切開で 2,812 g の女児を分娩し、Apgar スコアは 1 分値 5 点、5 分値 7 点であった。出生直後より人工呼吸管理を行い、軽度の肺高血圧を認めたが循環動態は安定していた。日齢 2 の CT 検査で右 CDH と診断し同日根治術を施行した。横隔膜背側で肝後区域が胸腔内に挙上し、腎臓は胸部の高さで後腹膜下に存在した。ヘルニア嚢に裂孔はなく有嚢性右 CDH と診断した。腎臓は腹部へ還納できなかった。横隔膜、腹横筋および Gerota 筋膜でヘルニア門の縫縮を行い日齢 46 に退院した。胸部異所性腎の胎児診断は稀だが、本症例は正中心がその契機となった。胸部異所性腎を合併した右 CDH では患側への縦隔偏位が起こりうることを認識した。

## Prenatal diagnosis of ectopic intrathoracic kidney with right congenital diaphragmatic hernia manifesting as fetal mesocardia

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## Abstract

Although an ectopic intrathoracic kidney can be complicated by congenital diaphragmatic hernia (CDH), prenatal diagnosis of this condition is rare. We report a rare case of prenatally diagnosed ectopic intrathoracic kidney with CDH presenting as fetal mesocardia. A 32-year-old gravida 1, para 0 showed fetal mesocardia on ultrasonographic screening performed at 18 weeks' gestation. No additional ultrasonographic anomalies were identified at the time. Detailed examination using magnetic resonance imaging at 30 weeks' gestation showed that the fetal right kidney and a part of the liver were intrathoracic in location, and we diagnosed right CDH. She underwent cesarean section at 37 weeks' gestation and delivered a female infant weighing 2,812 g with Apgar scores of 5-7. The infant was intubated immediately after birth, and mechanical ventilation was initiated. Although mild pulmonary hypertension was observed, her cardiorespiratory status stabilized smoothly. Computed tomography confirmed the diagnosis of right CDH, and we performed surgical repair on day 2 of birth. The posterior segment of the hepatic lobe and the right kidney were herniated through a right diaphragmatic defect and were covered by a hernia sac. The right kidney was located retroperitoneally and could not be replaced under the diaphragm. We diagnosed right CDH and concomitant ectopic intrathoracic kidney. We performed plication of the hernia sac with the transversus abdominis and Gerota's fascia. In this case, fetal mesocardia led to the prenatal diagnosis. Our case suggests that CDH can cause an ipsilateral mediastinal shift by complication with ectopic intrathoracic kidney.

## Keywords

ectopic intrathoracic kidney, congenital diaphragmatic hernia, mesocardia, prenatal diagnosis

## 1. はじめに

胸部は異所性腎が位置する部位として最も頻度の

低い部位であり、1 / 16,000 程度と報告されている<sup>1)</sup>。

胸部異所性腎は①背側横隔膜が正常な真の胸部異所性腎、②横隔膜弛緩症によるもの、③先天性横隔膜

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