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# A Rare Clinical Case of Prenatal Diagnosis of Isolated Right Lung Agenesis with Cardiac Dextraposition: Features of Differential Diagnosis

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**Annotation.** This article presents a rare case of isolated right lung agenesis with cardiac dextraposition in the fetus, interpreted as dextracardia at 22 weeks of pregnancy. The issues of differential diagnosis of isolated right lung agenesis with right heart displacement with dextracardia and other types of dextraposition are discussed.

**Keywords:** agenesis of the right lung, dextraposition of the heart, dextracardia, fetus, prenatal ultrasound diagnosis.

**Relevance.** Agenesis of the lung − this is a rare defect, the frequency of which is 1:15,000 births [1]. Isolated unilateral lung agenesis can be successfully diagnosed as early as the end of the first trimester of pregnancy [2]. The diagnosis of isolated lung agenesis is based on the absence of visualization on one side of the pulmonary artery and lung tissue. Isolated agenesis of the right lung leads to a displacement of the heart to the affected side and there is a need for differential diagnosis of various conditions when the fetal heart is located in the right half of the chest. This article demonstrates a clinical case of agenesis of the right lung with a shift of the heart to the right and discusses aspects of differential diagnosis in the prenatal period.

**Clinical observation.** Patient Y. R. is 29 years old, this pregnancy is the second, one child is alive and well. During the screening ultrasound examination of the fetus at 22 weeks of pregnancy, a preliminary diagnosis of “dextracardia”was established. The patient decided to prolong the pregnancy, was observed at the place of residence. At 28 weeks of pregnancy, a fetal echocardiography was performed to clarify the diagnosis. During the ultrasound examination, one live fetus, male in the pelvic position, corresponding to 28 weeks and 5 days of pregnancy, was visualized in the uterine cavity. The left lung was determined by the usual average echogenicity, occupied the left half of the chest cavity, the right lung was not visualized. The heart occupied the entire right chest, the Cardiothoracic index (CTO) was 0.67, the cardiofemoral index (CFI) was 0.55. The thoracic aorta was located near the spine along the median line. The axis of the heart lay parallel to the median line, the tip of the heart was facing forward (fig. 1). Morphologically, the right ventricle was located on the right. There was a concordant atrioventricular and ventriculoarterial junction. The output paths had an intersection (fig. 2). On the section through three vessels, the aorta was shifted to the right, the pulmonary trunk to the right and forward (fig. 3). There was a bifurcation of the pulmonary trunk with the departure of the left pulmonary artery and the ductus arteriosus. The right pulmonary artery was not visualized. The trachea was located to the left of the spine, the ductus arteriosus and the aortic arch were located to the right of it. The descending aorta at the level of the four-chamber section was located to the left of the spine. On a cross-section through the upper floor of the abdominal cavity, the stomach was located to the left of the spine (fig. 4). Conclusion. Agenesis of the right lung. Dextraposition of the heart. In the full term, a newborn boy weighing 2800, 47 cm tall, was born on the Apgar scale by 7-8 points. At the

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moment, the child develops according to age, he is 5 months old.

**Discussion.** Agenesis of the right lung with a shift of the heart to the right (dextraposition of the heart) must be differentiated from dextracardia.Dextracardia with the reverse arrangement of the abdominal organs (situs inversus totalis) is an anomaly in which there is a complete inversion of the internal organs with a "mirror" arrangement. It is formed in the period up to the 3rd week of embryonic development and is associated with a violation of the formation and spatial movement of the bulbar-ventricular loop. With this anomaly, the heart is located in the right half of the chest, the apex of the heart is directed to the right (right-placed heart), the venous atrium and the superior vena cava are located on the left (left-formed heart). There is also an inversion of the tracheobronchial tree with two lobes on the right and three lobes of the lungs on the left. The liver is located on the left in the abdominal cavity, the stomach and spleen are on the right [3].

In isolated dextrocardia, the heart is also located in the right half of the chest cavity, but the abdominal organs have a normal location. There is a concordant atrio-ventricular and ventricular-arterial junction. At the same time, the viscero-atrial correspondence is preserved: both the venous atrium and the liver are located on the right [3]. The tip of the heart is directed to the right −the right-placed, right-formed heart. For the dextraposition of the heart, due to the agenesis of the right lung, the same position of the heart is characteristic −right-placed, right-formed. The distinctive features of these two nasologies are the absence/presence of lung tissue on the right and the direction of the apex of the heart: in dextracardia, the apex is directed to the right, and in dextraposition of the heart, its axis lies parallel to the median line and the apex is facing forward. It should be noted that there are reports in the literature when, in isolated agenesis of the right lung and dextraposition of the heart, the apex of the heart was turned to the right [1].

Dextraposition of the heart, in addition to agenesis of the right lung, in the antenatal period, can be caused by left-sided diaphralmal hernia with displacement of the abdominal organs into the chest, congenital cystic-adenomatous malformation of the lung, bronchogenic, enterogenic cysts, atresia of the left main bronchus and hydrothorax on the left [4,5]. Their differential diagnosis is not difficult. In all of the above cases, ultrasound examination of the fetus finds pathognomonic signs inherent only in this defect. Diagnosis of hydrothorax is based on the detection of anechoic pleural effusion on the left [6]. In the case of diaphragmatic flu, the stomach or part of the intestine is found in the left chest cavity [7], with enterogenic cysts − a cystic formation in the projection of the left lung, while the stomach is located in the abdominal cavity [8]. Congenital cystic-adenomatous lung malformation, depending on the type of lesion, is characterized by the presence of large or multiple small cysts in the projection of the lung. The affected lung is enlarged in volume, with increased echogenicity. It is extremely important to adequately assess the echogenicity of the lung and its size during ultrasound examination. The third type of cystic-adenomatous lung defect deserves attention, in which the cysts are microscopic and they are not visualized during ultrasound examination [4,9]. Differential diagnostic criteria for this defect can be an increase in the size of the affected left lung with a pronounced increase in its echogenicity, as well as the presence of a hypoplasized right lung. The periodical literature also describes cases of isolated hypoplasia of the right lung with dextraposition of the heart [10]. In cases of agenesis of the right lung, observations of the erroneous representation of the compensatory enlarged left lung as congenital emphysema are described [11].

It should be noted that in isolated lung agenesis, the CPI indicators will be the most reliable for assessing the size of the heart, since due to a decrease in the size of the chest, the CTO indicators will be overestimated. In our clinical example, the heart is visually enlarged, occupies half of the chest, but nevertheless, given that the KFI index of 0.55 is within the normal range,

cardiomegaly is excluded in this case.

Thus, the above clinical observation is of particular interest to specialists of ultrasound diagnostics as a rare congenital heart defect and emphasizes the need for a differential diagnosis between the displacement of the heart to the right due to the agenesis of the right lung and the dextraposition of the heart of another etiology, as well as isolated dextracardia.

Agenesis of the right lung and the resulting dextraposition of the heart is characterized by the absence of lung tissue and the direction of the apex of the heart forward. Also, this defect is characterized by a change in the location of the main arteries on the section through three vessels that move to the right and forward. And in dextracardia, the tip of the heart is turned to the right, while the lung tissue on the right is determined.

**References:**

1. Гусева О.И., Галягина Н.А. Пренатальная диагностика агенезии правого легкого. Пренатальная диагностика. 2014;13 (3): 218–222.
2. Потолова Е.В., Кучева Е.Л. Случай пренатальной диагностики агенезии правого легкого в комплексе множественных врожденных пороков развития у плода в I триместре беременности. Пренатальная диагностика. 2016;15 (3): 253–256.
3. Маслякина А.А., Емельянова И.А., Плотко И.С. Опыт пренатальной диагностики декстрокардий. Ультразвуковая и функциональная диагностика. 2014; 6: 71-76.
4. Durell J., Ringo Y., Lakhoo K. Congenital Cystic Lung Lesions. Pediatric Surgery. 2020; 495-501. https://doi.org/10.1007/978-3-030-41724-6-44
5. Медведев М.В. Пренатальнаяэхография: дифференциальный диагноз и прогноз: Руководство для врачей ультразвуковой диагностики, специалистов по пренатальной диагностике, акушеров-гинекологов, перинатологов, генетиков / М. В. Медведев. – 4-е издание. – М.: Реал-Тайм. – 2016. – 640 с.
6. Deurloo K. L., Devlieger R., Lopriore E., Klumper F. J., Oepkes D. Isolated fetal hydrothorax with hydrops: a systematic review of prenatal treatment options. PrenatDiagn 2007; 27: 893–899.
7. Pelizzo G., Costanzo S., Selvaggio G.G.O., Rebosio F., Canazza L., Marinoni F.,Calcaterra V.Non-casual association between congenital pulmonary airway malformations/primary lung hypoplasia and congenital diaphragmatic hernia (CDH). Front. Pediatr. 8:446. https://doi: 10.3389/fped.2020.00446
8. Robles B.N., Hammas M., Saavedra C., Russell K., Faustin D. Prenatal diagnosis of a solitary pulmonary cyst in a male fetus. Journal of Pediatrics and Neonatal Medicine. 2020; 2 (1): 115-119.
9. Duncombe G. J., Dickinson J.E., Kikiros C. S. Prenatal diagnosis and management of congenital cystic adenomatoid malformation of the lung. Am J Obstet Gynecol. 2002; 187 (4): 950-954.
10. Abdullah M.M., Lacro R.V., SmallhornJ.,Chitayat D., van der Velde M.E., Yoo S.J., Oman-Ganes L.,Hornberger L.K. Fetal cardiac dextroposition in the absence of an intrathoracic mass: sign of significant right lung hypoplasia. J Ultrasound Med. 2000;19:669–676.
11. Ganescu O., Petroze R.T., Puligandla P.S. Unilateral Lung Agenesis in an infant with prenatal diagnosis of Congenital Lobar Emphysema. Journal of Pediatric Surgery Case Reports. 2020; 53: 1-3.

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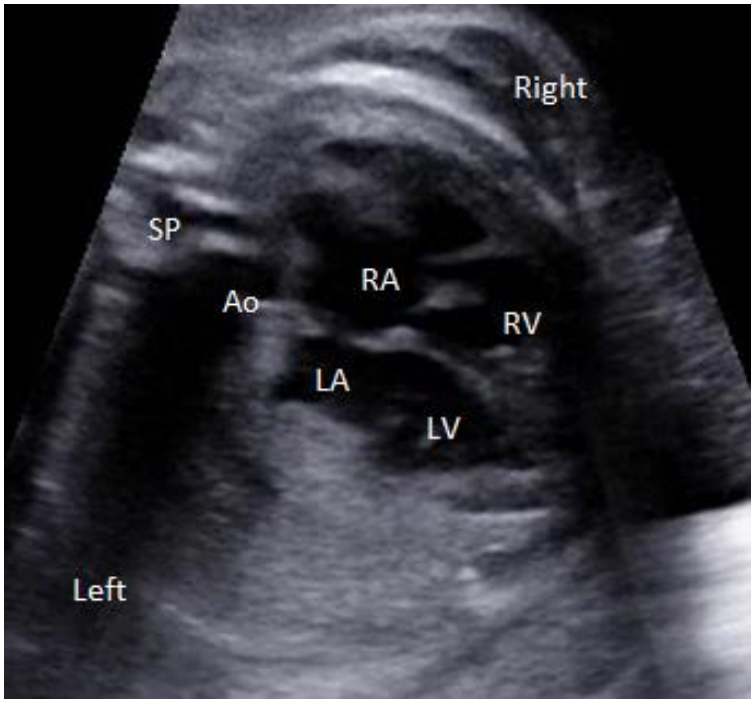


**Figure 1.** Agenesis of the right lung. Dextraposition of the heart. Pelvic attachment of the fetus. Four-chamber section of the heart. KFI 0.55. The axis of the heart is parallel to the median line, the apex of the heart is facing forward. Ao- aortic; SP-spinal; LA-left atrium; RA-right atrium; LV-left ventricle; RV-right ventricle.

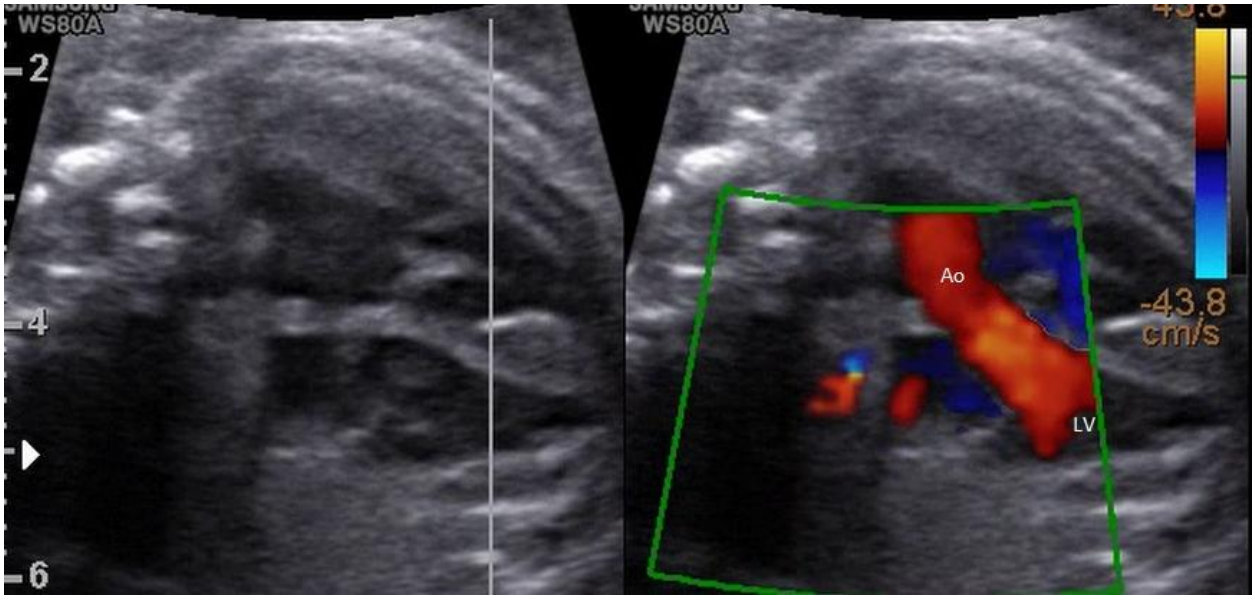


**Figure 2.** The left output path. The aorta exits the left ventricle. Pelvic attachment of the fetus. Ao- aortic; LV-left ventricle.

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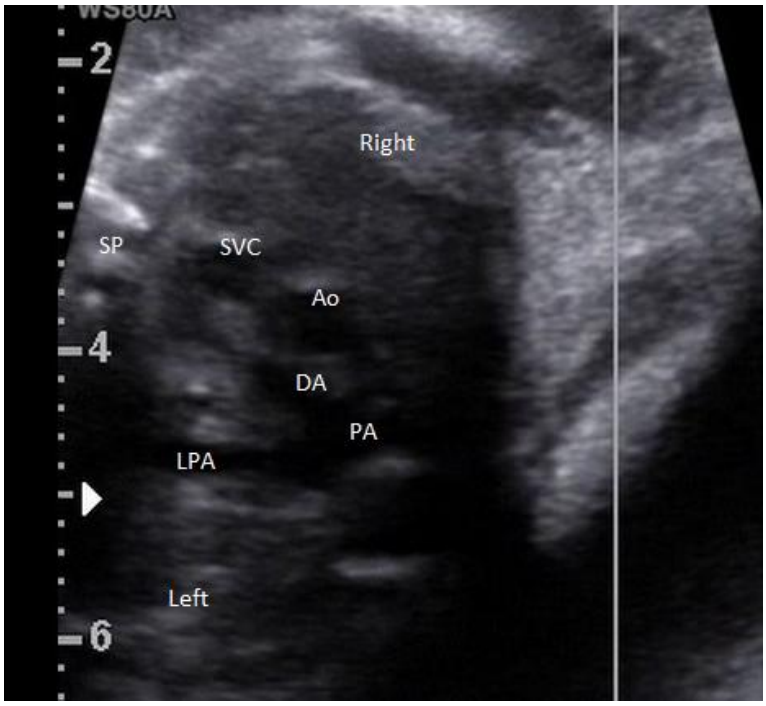


**Figure 3.** Cut through three vessels and the trachea. Pelvic attachment of the fetus. The ascending aorta is displaced to the right, the pulmonary trunk is displaced to the right and forward. The section shows the bifurcation of the pulmonary trunk into the ductus arteriosus and the left pulmonary artery. Ao-aortic; SP-; PA-pulmonary artery; LPA-left pulmonary artery; DA-ductus arteriosus; SVC-superior vena cava.

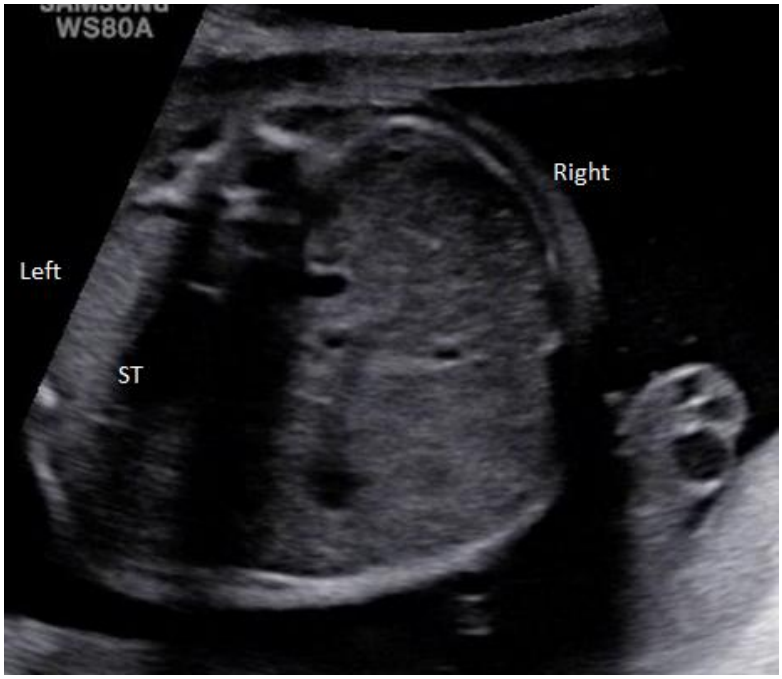


**Figure 4.** Cut through the upper floor of the fetal abdomen. Pelvic attachment of the fetus. The stomach is located on the left side of the abdomen. ST-stomach.