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КЛИНИЧЕСКИЙ СЛУЧАЙ: РЕДКИЙ СОЧЕТАННЫЙ ПОРОК РАЗВИТИЯ «ПОДКОВООБРАЗНОЕ ЛЕГКОЕ», СЕКВЕСТРАЦИЯ ЛЕГКОГО И СИНДРОМ ГИПОПЛАЗИИ ЛЕВЫХ КАМЕР СЕРДЦА

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РЕЗЮМЕ

Цель исследования. Обсудить возможности компьютерной томографии с контрастированием у новорожденных с подозрением на врожденные пороки развития сердечно-сосудистой системы и легких. **Материалы и методы.** Представлено клиническое наблюдение новорожденной с редким сочетанием подковообразного легкого с синдромом гипоплазии левых отделов сердца, гипоплазией и интралобарной секвестрацией правого легкого, аномалией системно венозного возврата, аномалией впадения печеночных вен. **Результаты.** Компьютерная томография с внутривенным контрастированием позволяет за одно исследование визуализировать анатомию сложного врожденного порока сердца, подтвердить наличие бронхолегочной аномалии. **Обсуждение.** Подковообразное легкое — это редкая врожденная аномалия, при которой имеется паренхиматозный перешеек между нижними отделами легких, располагающийся кпереди от аорты и позади левого желудочка сердца, содержащий бронхиальные и сосудистые структуры. Эта аномалия впервые была описана в 1962 году. И всего порядка 50 случаев во всем мире были зарегистрированы в литературе. В большинстве описанных случаев подковообразное легкое сочетается с синдромом ятагана или венолобарным синдромом. Синдром ятагана характеризуется аномальным венозным возвратом из правого легкого в нижнюю полую вену. **Заключение.** Данное клиническое наблюдение демонстрирует возможности постнатального обследования (КТ, прямая ангиография) в распознавании редчайшего порока развития легких и сердца — гипоплазии левых отделов сердца с гипоплазией и интралобарной секвестрацией правого легкого, аномалией системно венозного возврата, аномалией впадения печеночных вен, частота встречаемости которого составляет 1–3 случая на 100 000 новорожденных.

Ключевые слова: компьютерная томография, подковообразное легкое, секвестрация легкого, синдром гипоплазии левых отделов сердца.

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A RARE CONSTELLATION OF HORSESHOE LUNG WITH LUNG MALFORMATION AND HYPOPLASTIC LEFT HEART SYNDROME: A CASE REPORT

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ABSTRACT

Introduction. The aim of the paper is demonstrating the possibilities of CT angiography in newborns with suspected combinations of a congenital heart defect and pulmonary malformations. **Materials and methods.** The article presents a clinical case of a rare constellation of horseshoe lung with hypoplastic left heart syndrome, right lung hypoplasia and intralobar pulmonary sequestration of the right lung. **Results.** Contrast-enhanced computed tomography is mandatory in visualization of rare complex congenital heart and bronchovascular anomaly. **Discussion.** Horseshoe lung is a rare congenital anomaly of childhood in which the caudal and basal segments of the lungs are joined together anterior to the aorta and behind the left ventricle. This anomaly was described for the first time by Spenser in 1962. Horseshoe lung is often associated with unilateral lung hypoplasia, most commonly involving the right lung, and can occur in conjunction with scimitar syndrome, which includes hypoplasia of the right lung, abnormal right pulmonary venous return, and abnormal arterial supply to the right lung. **Conclusion.** Presented case report demonstrates current postnatal examination possibilities (CT, angiography) in the diagnostics of a rare congenital heart and bronchovascular anomaly — hyperplastic left heart, intralobar pulmonary sequestration of the right lung, anomalous

venous return, anomalous hepatic venous drainage. The incidence of this disorder is approximately 1–3 per 100 000 births.

Key words: congenital heart defect, congenital lung malformation, horseshoe lung, hypoplasia, intralobar sequestration.

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INTRODUCTION

Horseshoe lung is a rare congenital anomaly of childhood in which the caudal and basal segments of the lungs are joined together anterior to the aorta and behind the left ventricle (Fig. 1). This anomaly was described for the first time by Spenser in 1962 [1]. Horseshoe lung is often associated with unilateral lung hypoplasia, most commonly involving the right lung, and can occur in conjunction with scimitar syndrome, which includes hypoplasia of the right lung, abnormal right pulmonary venous return, and abnormal arterial supply to the right lung [2]. Horseshoe lung may also be diagnosed in very rare cases in combination with intralobar pulmonary sequestration, i.e. the situation in which a segment of lung parenchyma is not connected to the tracheobronchial tree. Other associations have also been reported, including various congenital heart defects, or foregut anomalies, that is why clinicians should be aware of associated birth defects and perform proper diagnostic evaluation in each case [3].

CASE REPORT

We report a rare case of a constellation of horseshoe lung with hypoplastic left heart syndrome, right lung hypoplasia and intralobar pulmonary sequestration of the right lung.

A full-term girl was born with a body weight of 3 170 g and Apgar score of 7/8. Her mother was diagnosed with gestational diabetes. The prenatal US-diagnosis of a fetal congenital heart defect was made: a hypoplastic left heart syndrome, multiple ventricular septal defects, single atrium, an anomaly of the systemic venous return. A single umbilical artery was also revealed. The diagnosis of a ductus-dependent congenital heart defect was confirmed on echocardiography after birth: hypoplastic left heart syndrome (hypoplasia of the left ventricle, hypoplasia of the mitral and aortic valves, hypoplastic aortic arch, coarctation of the aorta (fig. 2 a, b), ventricular septal defect. The patient was examined by a cardiologist and prescribed with prostaglandin infusion (Alprostan 30 ng/kg/min).

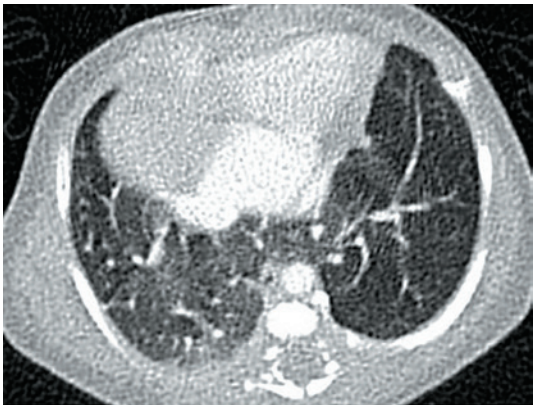
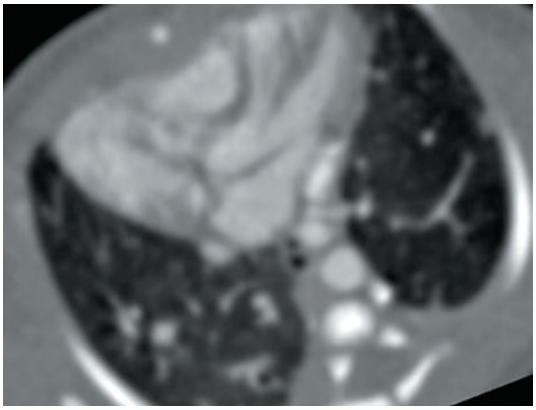


Figure 1. Horseshoe lung. A CT scan (axial view; lung window) shows an isthmus of the pulmonary tissue between the left and right lungs located to the front of the aorta and behind the left atrium and containing bronchi and vessels

Contrast-enhanced chest CT scan was performed on the 2 day after birth using a 128 slice CT scanner (Siemens Somatom Definition) demonstrating a congenital lung malformation in the form of horseshoe lung (fig. 3a, b) in combination of right lung hypoplasia and pulmonary artery hypoplasia (fig. 4a).

In addition, intralobar sequestration involving the entire right lower and right middle lobe was revealed

with a large aberrant artery of 2.5 mm in diameter branching from the abdominal aorta proximal to the celiac trunk (fig. 4c). Pulmonary angiography confirmed CT findings and showed that the left branch of the pulmonary artery was up to 3–3.5 in diameter and supplied all left lung. The right lobar pulmonary artery was 2 mm in diameter and supplied the right upper lobe, while the right lower and middle lobar arteries were supplied by a large vessel of 3.5 cm in diameter branching from the abdominal aorta at the level of the celiac trunk (fig 5a, b). The pulmonary sequester had its own vein draining into the atrium.



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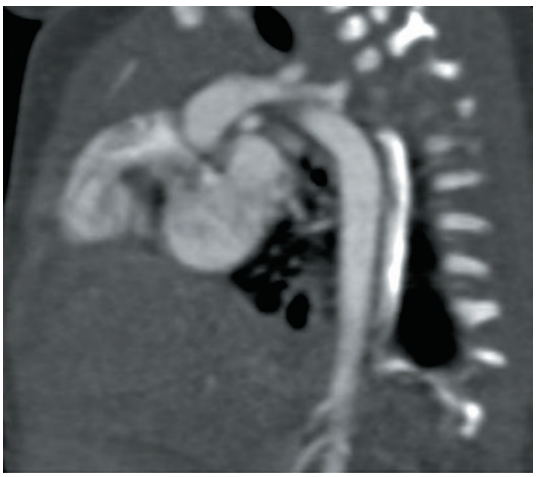

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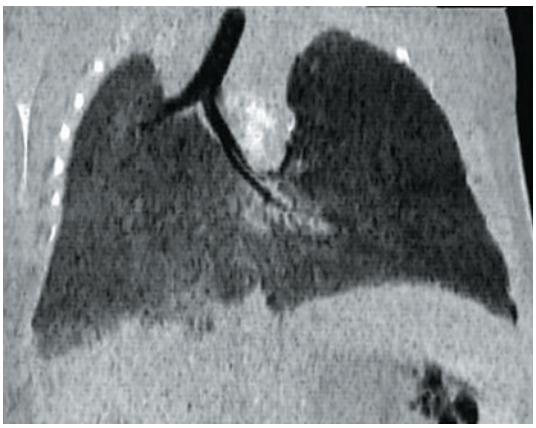
Figure 2. CT scans (axial and sagittal views, soft tissue window, MIP reconstruction technique). Hypoplastic left heart chambers (left ventricle and atrium are undersized, mitral valve hypoplasia) are visualized (a). Preductal coarctation of the aorta, patent ductus arteriosus and contrast enhancement of the descending aorta as a result of a ductal-dependent congenital heart defect (b)

Due to worsening of the child's condition, she was put on mechanical ventilation on the sixth day of life. On the tenth day of life, based on clinical and diagnostic data, the Norwood procedure with ligation of the vessels of the intralobar sequestration was performed. However, during the postoperative period the child's condition was unstable because of severe myocardial insufficiency while on therapy with subtoxic doses of inotropic agents and vasopressors, and she died on the eleventh day of life.



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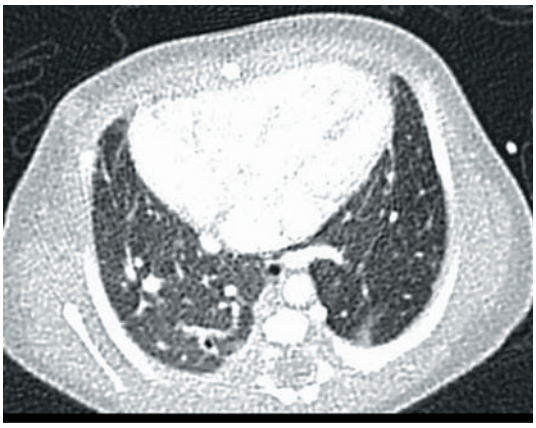


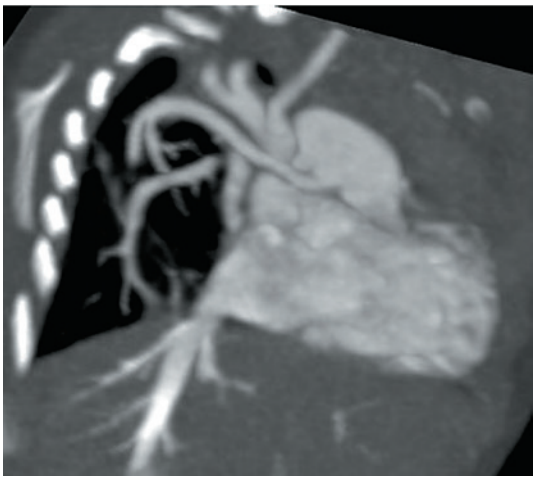
b

Figure 3. CT scans (axial view, lung window, MinIP reconstruction technique). Horseshoe lung with deviation of the tracheal bifurcation to the right due to the right lung hypoplasia (a). The left main bronchus passes through the parenchymal isthmus. The isthmus of lung tissue is situated at the level of the lower pulmonary regions and contains vessels and bronchi (b)

DISCUSSION

Constellations of various congenital malformations of the heart and the lungs is most likely a result of prenatal exposure to certain unfavorable factors during the early stage of pregnancy (about 3–4 weeks of gestation) [4]. This exposure results

in impaired development of the bronchial tree leading to pulmonary sequestration, and congenital heart defects. The results of animal studies suggest that some congenital malformations of the lung may be due to specific gene mutations as lung morphogenesis is regulated by various hormones, growth factors, and transcription factors [5]. In most of the case reports published, horseshoe lung was associated with right lung hypoplasia [6], and scimitar syndrome [7]. In our case, the girl had anomalies of the systemic venous return. From an accessory hemiazygos vein, the left-sided accessory superior vena cava, and the vein from the left liver lobe her blood drained into a dilated coronary sinus (Fig. 4b). The anatomy of the pulmonary veins was abnormal, and there was a single collector behind the posterior atrial wall so that the blood got into the common atrium. Another specific feature of our case was that hypoplastic left heart syndrome was accompanied by concomitant lung malformations resulting in severe respiratory failure and a higher potential risk for infections due to intralobar sequestration.



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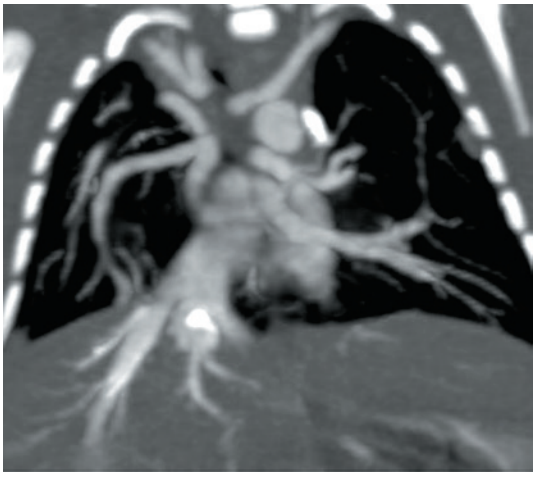

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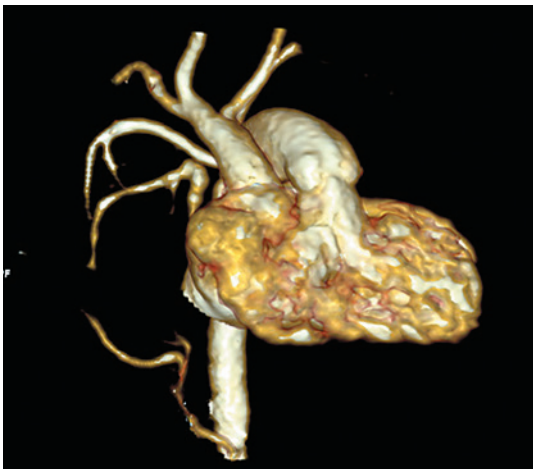


c

Figure 4. CT scans (a, b — soft tissue window, MIP reconstruction technique; c — Volume Rendering Technique). Hypoplastic right pulmonary artery, which has an anomalous pathway, supplies the right upper lobe (a, b). Anomalous pulmonary venous drainage is seen (b) with an accessory vessel branching from the abdominal aorta and supplying the right lung sequestration (c)

CONCLUSION

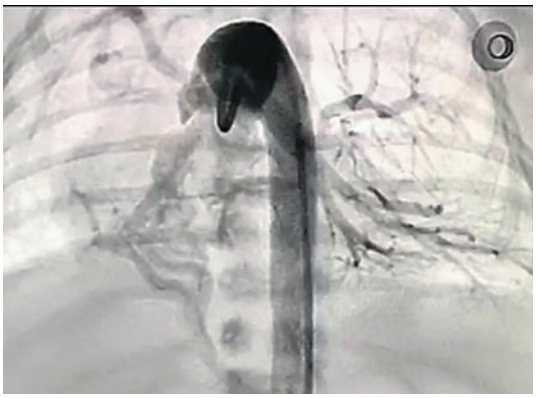
In newborns with suspected combinations of a congenital heart defect and pulmonary malformations, contrast-enhanced *chest and upper abdominal* CT angiography is appropriate imaging modality as it permits multiplanar reformatting and three-dimensional reconstructions, and allows obtaining information about potential congenital vascular anomalies involving the thoracoabdominal aorta and major veins. During the systemic arterial phase, special attention should be paid to the presence of:

1) anomalous vessels branching from the descending aorta or the proximal abdominal aorta, which might indicate a pulmonary sequestration.

In additional to the arterial phase, the diagnostic imaging algorithm in such cases should include the use of the venous *phase of contrast-enhanced-computed-tomography of the chest and upper abdomen* to assess the presence of:

1) anomalous systemic venous return;
1) partial anomalous pulmonary venous return;
2) vascular anomalies of the hepatic veins.

The use of the proper imaging strategy will ensure urgent diagnosis and timely surgical treatment in neonatal patients with rare combinations of congenital cardiac and pulmonary defects.



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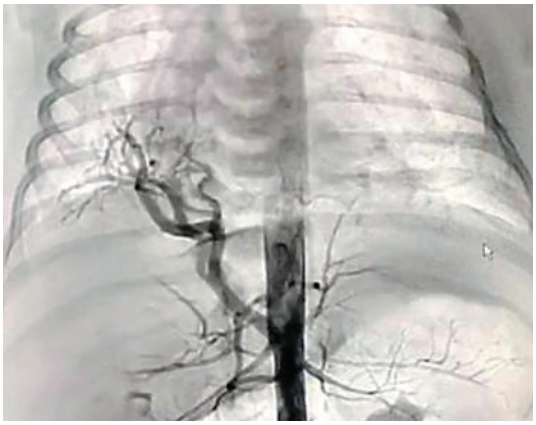

b

Figure 5. Pulmonary angiograms demonstrate a hypoplastic right pulmonary artery supplying the right upper lobe (a). A large vessel branching from the abdominal aorta and supplying the right lung sequestration is visualized (b)

Conflict of interest

The authors declare no conflict of interest.

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