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SCIMITAR SYNDROME IN AN INFANT IN THE CONTEXT OF MODERN IMAGING TECHNIQUES

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Scimitar syndrome is a rare congenital cardiopulmonary anomaly characterized by partial or complete anomalous drainage of the right pulmonary veins, hypoplasia of the right lung, and variable vascular abnormalities. The condition exhibits marked clinical polymorphism, ranging from severe manifestations in the neonatal period to an asymptomatic course with delayed diagnosis. This article presents a clinical case of an infant with scimitar syndrome associated with partial anomalous drainage of the right inferior pulmonary vein into the inferior vena cava, hypoplasia of the right pulmonary artery, systemic arterialization of the right lower lobe, and an atrial septal defect. The diagnostic value of multislice computed tomography angiography in delineating the anatomical features of the defect is emphasized. The clinical course, diagnostic considerations, and surgical management are discussed.

Key words: Scimitar syndrome, partial anomalous pulmonary venous return, pulmonary vascular anomalies.

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СИНДРОМ «ЯТАГАНА» У НЕМОВЛЯТИ З ТОЧКИ ЗОРУ СУЧАСНИХ МЕТОДІВ ВІЗУАЛІЗАЦІЇ

Синдром «ятагана» (Scimitar syndrome) – рідкісна вроджена кардіопульмональна аномалія, що характеризується частковим або повним аномальним дренажуванням правих легеневих вен, гіпоплазією правої легені та різними судинними аномаліями. Захворювання відрізняється вираженням клінічним поліморфізмом – від тяжких проявів у неонатальному періоді до безсимптомного перебігу з пізньою діагностикою. У статті представлено клінічний випадок синдрому «ятагана» у немовляти, що супроводжувався частковим аномальним дренажуванням правої нижньої легеневої вени в нижню порожнисту вену, гіпоплазією правої легеневої артерії, системним артеріальним кровопостачанням нижньої частки правої легені та дефектом міжпередсердної перегородки. Особливу увагу приділено діагностичній цінності мультиспіральної комп'ютерно-томографічної ангіографії для детального оцінювання анатомічних особливостей вади. Розглянуто особливості клінічного перебігу, діагностичні аспекти та хірургічну тактику.

Ключові слова: синдром «ятагана», частковий аномальний дренаж легеневих вен, аномалії легеневих судин.

Scimitar syndrome (also known as congenital venolobar syndrome) is one of the rarest and most diagnostically challenging combinations of cardiopulmonary anomalies, with an estimated incidence of 1–3 cases per 100,000 newborns [7]. The syndrome encompasses a heterogeneous spectrum of pulmonary venous, pulmonary arterial, bronchopulmonary, and systemic vascular abnormalities [5, 15]. The name derives from the characteristic radiographic appearance: on a frontal chest radiograph, a curved shadow running parallel to the right cardiac border resembles a Turkish sword, or scimitar. The constant components of Scimitar syndrome include hypoplasia (dysplasia) of the right lung and partial or complete anomalous pulmonary venous drainage (APVD) [15].

Partial anomalous pulmonary venous drainage (PAPVD) is characterized by the absence of connection of one or more pulmonary veins with the left atrium. The prevalence of PAPVD is approximately 0.3 % among all congenital heart defects (CHD) [6]. In these cases, the pulmonary veins may drain into the right atrium, the coronary sinus, the superior vena cava, or the inferior vena cava. Different combinations of anomalous drainage are classified as the mixed type.

The variable components include hypoplasia or aplasia of the pulmonary artery, systemic arterial supply to the lung, and pulmonary sequestration.

The syndrome was first described in 1836 based on the autopsy findings of an infant with dextroposition of the heart and hypoplasia of the right lung. Subsequently, it was described again in 1949 in an asymptomatic living patient using cardiac catheterization. The first documented surgical intervention was performed in 1950 in a patient with recurrent pneumonia, while the first corrective operation was carried out in 1956 [10]. However, it was not until 1960 that the syndrome was formally named, characterized, and correlated with radiological findings [14]. In 1973, B. Felson referred to this anomaly as congenital venolobar syndrome.

In cases where the descending right pulmonary vein follows an anomalous course within the right lung but drains normally into the left atrium, the condition is referred to as pseudo-Scimitar syndrome. In Scimitar syndrome, varying degrees of hypoplasia and dysplasia of the right lung are observed. An accessory tracheal bronchus, a rare malformation of the bronchopulmonary system, may also be present.

Hypoplasia or aplasia of the right pulmonary artery, as well as its localized stenoses, may occur. Narrowing of the anomalous vein at its junction with the inferior vena cava is possible, along with sequestration of pulmonary parenchyma resulting from anomalous systemic arterial supply to a portion of the right lung (most commonly the lower lobe) arising from the abdominal aorta. Dual pulmonary

blood supply combined with pulmonary venous obstruction may more rapidly lead to congestive heart failure and pulmonary arterial hypertension.

Scimitar syndrome may be associated with other congenital heart defects, including septal defects, patent ductus arteriosus, obstructive left heart lesions, tetralogy of Fallot, single ventricle physiology, Bland–White–Garland syndrome, double outlet right ventricle, and others [8, 10, 11].

Scimitar syndrome may present in the neonatal period with severe symptoms or remain undiagnosed for a prolonged time in cases with a mild clinical course. The frequency of respiratory infections depends, among other factors, on the degree of lung hypoplasia. Prenatal diagnosis may be suspected in the presence of cardiac dextroposition, right lung and right pulmonary artery hypoplasia, particularly when these findings are identified in the absence of diaphragmatic hernia [3].

Plain chest radiography may reveal the characteristic curvilinear vascular shadow along the right cardiac border. However, the sensitivity of this method is limited, particularly in newborns. Transthoracic echocardiography serves as the primary modality for evaluating intracardiac anatomy, pulmonary artery pressure, and detecting associated congenital anomalies. Direct visualization of anomalous pulmonary venous drainage is not always feasible.

In recent years, multislice computed tomography (CT) angiography and cardiovascular magnetic resonance imaging (MRI) have become key modalities for comprehensive anatomical assessment of Scimitar syndrome. They allow accurate delineation of anomalous pulmonary venous drainage, pulmonary arterial anatomy, systemic arterial supply, associated pulmonary abnormalities, and the spatial relationships between cardiovascular structures [2,12]. MRI additionally provides functional information on blood flow and shunt volumes without ionizing radiation, whereas CT angiography offers rapid acquisition and high spatial resolution, which may be particularly advantageous in infants [12].

In Scimitar syndrome, the following surgical interventions may be performed: 1) ligation and division of the anomalous vein with subsequent anastomosis to the left atrium; 2) conversion of the anomalous drainage to a cardiac form, followed by redirection of pulmonary venous return to the left atrium through an atrial septal defect using a pericardial patch; 3) creation of an intra-atrial tunnel between the orifice of the anomalous vein and the atrial septal defect, with enlargement of the inferior vena cava orifice using a patch when necessary. Contemporary surgical strategies continue to evolve, with modified techniques aimed at achieving unobstructed pulmonary venous drainage while minimizing the risk of postoperative venous stenosis [4, 6, 13].

In the late postoperative period, obstruction of a long intra-atrial tunnel may occur due to its complex anatomical course and angulation approaching 180°.

When the anomalous vein is located posterior to the root of the lung, its connection to the atrium may be technically unfeasible. In such cases, resection of the hypoplastic right lung or lung transplantation may be considered. Compensatory growth of the left lung can offset the absence of the right lung. [13]. The outcomes of surgical treatment depend on the patient's age, anatomical characteristics of the defect, associated anomalies, and the type of surgical procedure. Results are not always satisfactory, largely due to the potential development of pulmonary venous obstruction.

The purpose of the study was to establish Scimitar syndrome in an infant using modern imaging techniques.

Materials and methods. This publication presents a descriptive longitudinal clinical case of an infant with Scimitar syndrome who was examined and followed at the Scientific Research Institute of Pediatrics, Baku, Azerbaijan. The patient was initially hospitalized at the age of 3.5 months with respiratory symptoms and was subsequently reassessed at 9 months of age because of recurrent respiratory manifestations. Clinical follow-up continued until the age of 1 year and 1 month.

The diagnostic evaluation included collection of perinatal and postnatal history, comprehensive physical examination, routine laboratory investigations, chest radiography, transthoracic echocardiography, and multislice computed tomography (CT) angiography of the heart and great vessels. Clinical assessment included evaluation of the general condition, respiratory and cardiovascular status, cardiac and pulmonary auscultation, peripheral pulse characteristics, and signs of heart failure or respiratory compromise.

Chest radiography was used as the initial imaging modality for assessment of the pulmonary parenchyma, bronchovascular pattern, cardiac position and dimensions, and mediastinal structures. Particular attention was paid to the presence of an abnormal vascular shadow along the right cardiac border. Transthoracic echocardiography was subsequently performed to assess cardiac anatomy, atrioventricular and ventriculoarterial connections, cardiac chamber dimensions, ventricular systolic function, intracardiac shunts, valvular function, pulmonary arteries, and associated congenital heart defects. Doppler echocardiography was used for assessment of intracardiac blood flow and valvular regurgitation.

Multislice CT angiography of the heart and great vessels was performed for detailed characterization of pulmonary venous anatomy and associated cardiovascular and pulmonary abnormalities. The examination was aimed at determining the course and drainage site of the anomalous pulmonary vein, evaluating the pulmonary arteries and systemic arterial supply to the lungs, identifying associated intracardiac abnormalities, and assessing lung morphology and cardiac position. The final diagnosis was established

on the basis of combined clinical and multimodality imaging findings.

The therapeutic strategy was determined individually according to the patient's clinical condition, anatomical characteristics of the congenital defect, cardiac function, associated cardiovascular abnormalities, and potential indications for surgical correction. Following stabilization of the initial respiratory condition, the child remained under the supervision of a pediatric cardiologist and cardiac surgeon. At the latest follow-up, definitive surgical correction was being planned.

Due to the single-patient descriptive design, inferential statistical analysis was not applicable. Clinical and instrumental findings were analyzed descriptively and are presented as absolute values and imaging measurements.

The clinical observation was conducted in accordance with generally accepted principles of biomedical ethics and the Declaration of Helsinki. Written informed consent for the use and publication of anonymized clinical data and diagnostic images was obtained from the child's parent(s)/legal guardian(s).

Results of the study. Perinatal and neonatal history. Patient F., aged 3.5 months, was admitted in a moderately severe condition to the Research Institute of Pediatrics. At admission, the primary complaints were cough and lethargy. The child was born from the first pregnancy and first delivery at 37 weeks of gestation via spontaneous vaginal delivery. Birth was complicated by moderate asphyxia, and primary resuscitation was provided.

Clinical examination at hospitalization. Upon hospitalization, the patient demonstrated catarrhal symptoms, including pharyngeal hyperemia, nasal congestion, and low-grade fever. The skin appeared pale, and subcutaneous fat development was satisfactory. Auscultation of the lungs revealed diffuse moist rales. Performed investigations (routine clinical tests, neurosonography, and abdominal ultrasound) showed no pathological findings.

Radiographic findings. Chest radiography demonstrated no focal or infiltrative shadows in either lung. The bronchovascular pattern was enhanced in the perihilar regions bilaterally. The roots of both lungs were structured, pleural sinuses were free, and the upper mediastinal contours were mildly widened due to the thymus. The cardiac borders were expanded to the right.

After treatment, the child was discharged home in satisfactory condition. Periodically, the child experienced wheezing in the lungs and cough.

At 9 months of age, the child presented on an outpatient basis to the Research Institute of Pediatrics with complaints of catarrhal symptoms of the upper respiratory tract, cough, and moist rales in the lungs.

A chest X-ray revealed infiltrative focal shadows in both lungs. The bronchovascular pattern was enhanced in the perihilar regions on both sides. A broad vascular shadow was detected, running along the right border of the heart toward the diaphragm (Fig. 1).

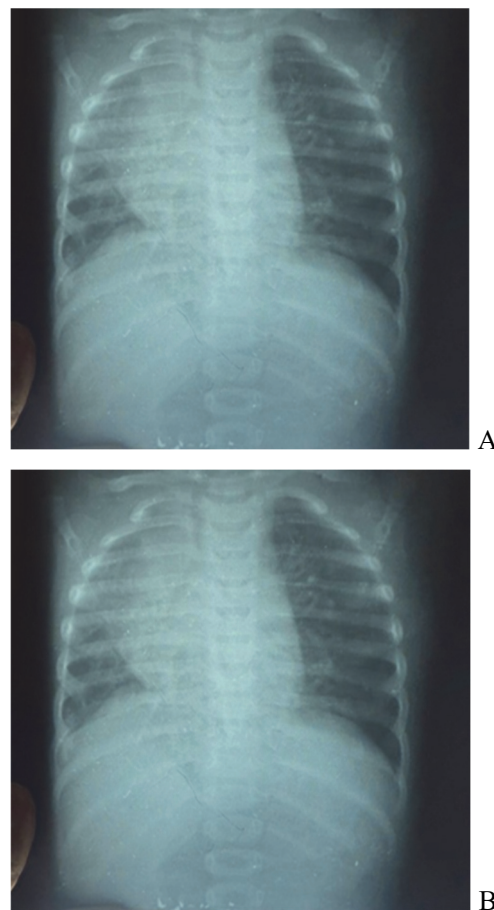


Fig. 1. A, B – The chest radiograph reveals a broad vascular shadow extending along the right cardiac border toward the diaphragm.

The recurrent respiratory manifestations together with the abnormal vascular shadow along the right cardiac border raised suspicion of an underlying congenital cardiopulmonary vascular anomaly. Therefore, the diagnostic strategy was expanded from routine respiratory assessment to targeted cardiovascular imaging. Transthoracic echocardiography was performed as the next diagnostic step to evaluate intracardiac anatomy, hemodynamics, and possible associated congenital heart defects.

Patient was referred to the Central Clinic for an echographic examination. On clinical examination, the general condition was satisfactory. Cardiac auscultation revealed a systolic murmur of grade 2/6 (Levine scale) in the mesocardial area. Femoral and radial pulses were symmetric and adequately filled. To clarify the nature of the murmur, transthoracic echocardiography was performed.

Echocardiographic Findings. The cardiac apex was directed leftward. Atrial situs was solitus. Atrioventricular and ventriculoarterial connections were concordant. The relationship of the great vessels (aorta/pulmonary artery) was normal. Cardiac chambers were of normal size, with no evidence of dilation or hypoplasia.

An aberrant muscular trabecula (aberrant band) was visualized along the free wall of the left ventricle extending toward the interventricular septum. Myocardial contractility was preserved. Left

ventricular systolic function was satisfactory, with an ejection fraction (EF) of 70 %. Right ventricular contractility was normal.

A secundum atrial septal defect (ASD) with left-to-right shunting was identified at the level of the fossa ovalis. The mitral valve structure was normal, with no regurgitation. The tricuspid valve was structurally preserved, with mild tricuspid regurgitation (maximum velocity 2.1 m/s). No diastolic dysfunction was detected.

No ventricular septal defects were observed. The left ventricular outflow tract was unobstructed. The aortic valve was tricuspid, without stenosis or insufficiency (flow velocity 116 cm/s). The ascending aorta and aortic arch were normal, with no signs of coarctation. The right ventricular outflow tract and pulmonary valve were unobstructed (flow velocity 122 cm/s). The pulmonary trunk and branches were normally formed. Patent ductus arteriosus was not visualized. No pericardial effusion or intracardiac masses were detected. The myocardium showed no signs of noncompaction or pathological inclusions (Fig. 2).

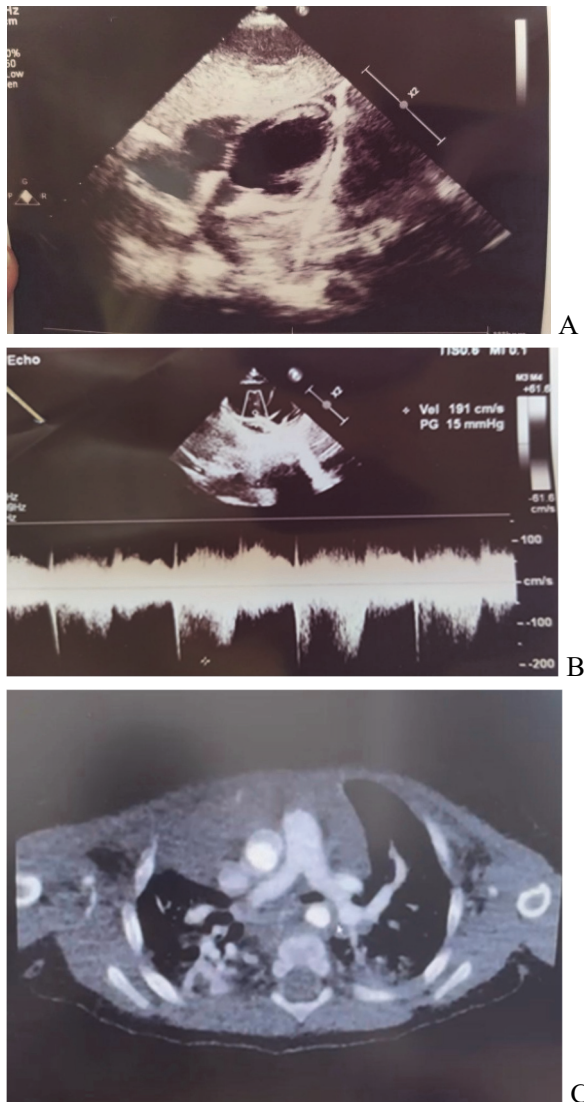


Fig. 2. A, B – Echocardiography; C – CT angiography.

The echocardiographic picture was consistent with a secundum atrial septal defect with left-to-right

shunting and mild tricuspid regurgitation. Myocardial contractile function was preserved.

Echocardiography demonstrated a secundum atrial septal defect with left-to-right shunting and mild tricuspid regurgitation, while biventricular systolic function remained preserved. However, echocardiography did not provide complete characterization of the pulmonary venous anatomy. Considering the abnormal radiographic vascular shadow and the need to exclude anomalous pulmonary venous drainage, CT angiography of the heart and great vessels was performed.

CT Angiography Findings. CT angiography of the heart and great vessels demonstrated situs solitus (abdominal, atrial, and tracheobronchial). Dextroposition of the heart with rightward mediastinal shift was noted, with the heart located predominantly in the right hemithorax. Cardiac configuration was characterized by mesocardia, D-looping, and preserved atrioventricular and ventriculoarterial concordance.

Cardiac chambers were balanced, without dilation or hypoplasia. The interventricular septum was intact. A secundum atrial septal defect (ASD II) measuring 4.5–5.0 mm was identified.

Partial anomalous pulmonary venous return (PAPVR) consistent with Scimitar syndrome was detected: the right inferior pulmonary vein (Scimitar vein) exhibited anomalous supradiaphragmatic drainage into the inferior vena cava. The Scimitar vein measured approximately 5.0 mm in diameter and followed a tortuous course, forming an anastomosis with the terminal portion of the right superior pulmonary vein. The left pulmonary veins and right superior pulmonary veins drained normally into the left atrium.

Additionally, anomalous systemic arterial supply to the lower lobe of the right lung was identified, arising from a branch of the proximal celiac trunk (diameter approximately 5.0 mm), consistent with systemic arterialization of the right lower pulmonary segment.

The aorta originated from the left ventricle, forming a left-sided arch with a three-vessel branching pattern. No evidence of coarctation, interrupted arch, aortic stenosis, or patent ductus arteriosus was observed. Coronary arteries demonstrated normal origin.

The pulmonary trunk and pulmonary arteries were confluent. The right pulmonary artery was hypoplastic relative to the left. The right lung was reduced in volume, and both lungs exhibited a mosaic ventilation pattern.

Integration of the clinical and multimodality imaging findings established the final diagnosis of Scimitar syndrome characterized by partial anomalous pulmonary venous return of the right inferior pulmonary vein into the inferior vena cava, hypoplasia of the right pulmonary artery, reduced right lung volume, anomalous systemic arterial supply to the right lower lobe, cardiac dextroposition, and an associated secundum atrial septal defect. Thus, CT angiography was decisive in defining the

complete anatomical spectrum of the congenital anomaly and provided the anatomical information required for further therapeutic and surgical planning.

Clinical diagnosis and early outcome. Following establishment of the diagnosis, the patient was referred for specialized pediatric cardiology and cardiac surgical follow-up. Because ventricular systolic function was preserved and no overt signs of congestive heart failure or acute hemodynamic decompensation were documented, emergency surgical intervention was not required during the reported observation period.

Management and follow-up. The management strategy therefore consisted of clinical and cardiological surveillance while the anatomical findings obtained by CT angiography were evaluated for definitive surgical planning. At the latest follow-up, at the age of 1 year and 1 month, the child remained under the supervision of a pediatric cardiologist and cardiac surgeon, and surgical correction was planned.

Discussion. Scimitar syndrome represents a rare and anatomically heterogeneous congenital cardiopulmonary malformation in which anomalous pulmonary venous drainage constitutes only one component of a broader spectrum of abnormalities involving the lung parenchyma, pulmonary arteries, systemic arterial supply, and cardiac position [7, 8, 15]. The marked variability of its anatomical substrate explains the considerable diversity of clinical presentations, ranging from severe respiratory or cardiovascular compromise during infancy to an incidental diagnosis later in life [9, 10, 15]. Recent pediatric data further emphasize that Scimitar syndrome should be regarded as a heterogeneous congenital condition requiring individualized anatomical and hemodynamic assessment rather than as an isolated anomalous pulmonary venous connection [5].

The present case illustrates several characteristic features of the syndrome and, at the same time, demonstrates the diagnostic difficulties that may arise during infancy. The patient initially presented at 3.5 months of age with predominantly respiratory manifestations, including cough, wheezing, and moist pulmonary rales, without clear clinical evidence of a complex cardiovascular anomaly. At 9 months, recurrent respiratory symptoms prompted repeated chest radiography, which demonstrated the characteristic broad vascular shadow extending along the right cardiac border toward the diaphragm. This clinical evolution is consistent with the known variability of Scimitar syndrome, particularly in pediatric patients, in whom recurrent respiratory symptoms may precede recognition of the underlying vascular anomaly [1, 5, 10].

An important feature of this case is the complementary role of different imaging modalities. Conventional chest radiography provided the first important clue by demonstrating the characteristic scimitar-like vascular shadow. Nevertheless, radiography alone cannot reliably define the origin,

course, or drainage site of the anomalous pulmonary vein or demonstrate the full spectrum of associated vascular abnormalities. Echocardiography subsequently identified a secundum atrial septal defect with left-to-right shunting and mild tricuspid regurgitation, while ventricular function remained preserved. However, complete characterization of the pulmonary venous anatomy required cross-sectional imaging. This observation supports the concept that echocardiography, although indispensable for assessment of intracardiac anatomy and hemodynamics, may need to be complemented by CT or MR imaging when anomalous pulmonary venous return is suspected [2, 12].

In the present patient, CT angiography was decisive for establishing the complete anatomical diagnosis. It demonstrated supradiaphragmatic drainage of the right inferior pulmonary vein into the inferior vena cava, hypoplasia of the right pulmonary artery, reduced right lung volume, and anomalous systemic arterial supply to the right lower lobe arising from the proximal celiac trunk. These findings illustrate the particular strength of CT angiography in Scimitar syndrome, as it enables simultaneous assessment of pulmonary venous drainage, pulmonary arterial anatomy, systemic collateral vessels, lung morphology, and their three-dimensional anatomical relationships [2, 12]. Masrani et al. specifically emphasized the value of CT and MR imaging in demonstrating the anatomical associations and radiological characteristics of this syndrome. Such detailed anatomical mapping is especially important when surgical or catheter-based intervention is being considered [12].

Cardiovascular MRI represents an important alternative or complementary technique because it avoids ionizing radiation and allows functional assessment of blood flow and shunt magnitude. Nevertheless, in infants, CT angiography may offer practical advantages when rapid acquisition and high spatial resolution are required. The choice of modality should therefore be individualized according to the patient's age, clinical stability, diagnostic question, and the need for detailed assessment of small vascular structures [2]. In our patient, CT angiography provided the anatomical information necessary to define not only the anomalous pulmonary venous connection but also the associated systemic arterialization and pulmonary arterial hypoplasia, findings that are directly relevant to subsequent therapeutic planning.

The combination of Scimitar syndrome with additional congenital cardiovascular abnormalities is well recognized. Associated lesions may include atrial or ventricular septal defects, patent ductus arteriosus, and more complex congenital heart disease [8, 11, 15]. In the present case, a secundum atrial septal defect accompanied the anomalous pulmonary venous return. The presence of an interatrial communication may increase left-to-right shunting and therefore has potential implications for pulmonary hemodynamics and the timing of intervention. The coexistence of right pulmonary

artery hypoplasia and systemic arterial supply to the right lower lobe further illustrates the complex vascular phenotype of the syndrome.

The age at presentation is clinically important. Severe infantile forms may be associated with respiratory distress, pulmonary hypertension, heart failure, pulmonary venous obstruction, and significant associated congenital heart disease [1, 15]. Conversely, milder variants can remain clinically silent until adulthood [9]. Prenatal recognition is also possible and may facilitate early postnatal evaluation, particularly when fetal imaging demonstrates cardiac dextroposition, right lung hypoplasia, abnormal pulmonary vascular anatomy, or associated congenital heart disease [3]. The absence of prenatal recognition in the present case and the initially nonspecific respiratory manifestations demonstrate why a high index of suspicion remains necessary in infants with recurrent respiratory symptoms and unusual radiographic findings.

Management of Scimitar syndrome should be individualized according to symptoms, shunt magnitude, pulmonary artery pressure, pulmonary venous obstruction, systemic arterial supply, associated cardiac defects, and the anatomical characteristics of the anomalous vein [6, 13, 15]. Surgical treatment is primarily directed toward establishing unobstructed pulmonary venous return to

the left atrium and correcting clinically significant associated lesions. Various techniques have been described, including direct reimplantation and intracardiac baffling, while contemporary modifications continue to be developed to reduce the risk of postoperative pulmonary venous obstruction [4, 6]. Long-term outcome is strongly influenced by the complexity of the anatomy and by the development of pulmonary venous stenosis or obstruction after intervention [13, 15].

In the presented patient, preserved ventricular function and the absence of overt heart failure allowed continued cardiological and cardiac surgical surveillance while definitive correction was being planned. Nevertheless, the coexistence of anomalous pulmonary venous drainage, systemic arterialization of the right lower lobe, right pulmonary artery hypoplasia, and an atrial septal defect requires careful longitudinal hemodynamic assessment. This case therefore reinforces the importance of a multimodality diagnostic strategy in Scimitar syndrome. While radiography and echocardiography remain essential initial investigations, CT angiography can provide a comprehensive anatomical roadmap and may play a decisive role in diagnosis and preoperative planning, particularly in anatomically complex pediatric cases [5, 12].

Conclusion

Scimitar syndrome is a complex congenital anomaly with variable anatomy and clinical manifestations. The presented clinical case highlights the need for comprehensive instrumental evaluation in patients suspected of pulmonary venous return anomalies. Although chest radiography and echocardiography may have limited sensitivity in certain cases, multislice CT angiography allows detailed visualization of vascular anatomy, identification of systemic arterial supply, and assessment of pulmonary parenchyma.

Early diagnosis of the syndrome is crucial for selecting the optimal surgical strategy and for preventing the development of pulmonary hypertension and heart failure. The prognosis depends on the anatomical characteristics of the defect, the severity of hemodynamic disturbances, and the timeliness of corrective intervention.

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CLINICAL CASE OF APLASIA CUTIS CONGENITA HIGHLIGHTING THE DIAGNOSTIC SIGNIFICANCE OF THE HAIR COLLAR SIGN IN A NEWBORN

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Aplasia cutis congenita is a rare congenital disorder characterized by the absence of skin at birth, most commonly affecting the scalp. The aim of the study is to present a clinical observation of a case of congenital aplasia of the skin in a child. The article describes the case of girl K., hospitalized on the 4th day for examination and determination of management tactics due to the presence of 2 blisters filled with clear fluid in the parietal area, which were determined from birth. The skin around them was not changed, along the head to the right of the sagittal suture there was a strand of long dark hair. At 3 months of life one of the blisters has become less tense, its volume has decreased, a white scar begins to form in the center. At 6 months – it was observed that atrophic flat scars covered with thin epidermis formed in place of both bullae. ACC is rare and therefore the awareness of neonatologists, pediatricians and dermatologists is extremely important for early clinical diagnosis and conducting studies to exclude a concomitant brain defect, especially in the presence of a "hair collar" symptom, in order to prevent further complications.

Key words: aplasia cutis congenita, newborn, blisters, hair collar symptom.

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КЛІНІЧНИЙ ВИПАДОК ВРОДЖЕНОЇ АПЛАЗІЇ ШКІРИ У НОВОНАРОДЖЕНОГО З АКЦЕНТОМ НА ДІАГНОСТИЧНЕ ЗНАЧЕННЯ СИМПТОМУ ВОЛОСЯНОГО КОМІРЦЯ

Вроджена аплазія шкіри – це рідкісне вроджене захворювання, що характеризується відсутністю шкіри при народженні, найчастіше вражаючи волосисту частину голови. Метою дослідження є представлення клінічного спостереження за випадком вродженої аплазії шкіри у дитини. У статті описано випадок дівчинки К., госпіталізованої на 4-й день для обстеження та визначення тактики ведення у зв'язку з наявністю 2 пухирів, заповнених прозорою рідиною, утім'яній ділянці, які визначалися з народження. Шкіра навколо них не змінена, вздовж голови праворуч від сагітального шва було пасмо довгого темного волосся. На 3 місяці життя один з пухирів став менш напруженим, його об'єм зменшився, у центрі почав формуватися білий рубець. На 6 місяці життя було виявлено, що на місці обох булл утворилися атрофічні плоскі рубці, покриті тонким епідермісом. Вроджена аплазія шкіри зустрічається рідко, тому обізнаність неонатологів, педіатрів та дерматологів надзвичайно важлива для ранньої клінічної діагностики та проведення досліджень з метою виключення супутнього дефекту мозку, особливо за наявності симптому волосяного комірка, щоб запобігти подальшим ускладненням.

Ключові слова: вроджена аплазія шкіри, новонароджений, пухирі, симптом волосяного комірка.

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Aplasia cutis congenita (ACC) is a rare and clinically heterogeneous congenital disorder characterized by the complete or partial absence of skin at birth. The condition most frequently affects the scalp, although lesions may occur on virtually any anatomical site and may vary substantially in number, morphology, size, and depth of tissue involvement

[10]. ACC may present as a small superficial defect, an ulceration, a membranous lesion, or a fluid-filled/bullous lesion, while extensive forms may involve the subcutaneous tissues, calvarium, dura mater, and intracranial structures [2, 6]. The rarity of the disorder and its marked phenotypic variability may contribute to diagnostic difficulties, particularly