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RADIOLOGICAL METHODS IN THE DIAGNOSIS OF TOTAL ANOMALOUS PULMONARY VENOUS RETURN

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Abstract

Congenital heart defects (CHDs) account for one-third of all congenital malformations and occur in 0.7–1.7% of newborns. In the structure of CHD, the incidence of total anomalous pulmonary venous drainage is 1.1–3.0%.

In this scientific review, a comparative analysis of radiological research methods, such as echocardiography, catheterization of the heart cavities, magnetic resonance imaging and computed tomography, was carried out in the diagnosis and postoperative evaluation of the structures of total anomalous pulmonary venous drainage.

The list of literature consists of 40 publications, including one domestic and 39 foreign authors, based on meta-analysis, systematic reviews and clinical studies.

Total anomalous pulmonary venous return (TAPVC) is associated with high mortality. Life expectancy in patients with a non-obstructive form of TAPVC is higher than obstructive. In the non-obstructive form of TAPVC, 50% mortality is noted by three months of life and 80% mortality by 12 months. Patients with obstructive TAPVC die in the neonatal period. Newborns with TAPVC have an unfavorable prognosis of the disease without surgical treatment. Only a fifth survive to the age of 1 year. In half of the patients who died in the first three months of life, a lethal outcome was observed in the first week after birth. Early diagnosis and an accurate anatomical picture of this type of defect play an important role in the subsequent phased surgical correction. Echo in this regard was insufficient to visualize individual types of TAPVC. Cardiac catheterization, unlike CT, is associated with a relatively high risk of developing immediate and long-term complications.

Echocardiography is a routine method for the primary diagnosis of TAPVC. CT, in turn, is the "gold standard" and the method of choice for adequate pre-and postoperative diagnosis of the defect. When CT is not possible, and echocardiography is limited, MRI is an excellent method of choice. Cardiac catheterization is advisable only for specific indications, preferably for children older than a year because of the development of serious complications.

Key words: *total anomalous pulmonary venous return diagnostics, echocardiography, CT, catheterization, ionizing radiation.*

Резюме

РАДИОЛОГИЧЕСКИЕ МЕТОДЫ ИССЛЕДОВАНИЯ В ДИАГНОСТИКЕ ТОТАЛЬНОГО АНОМАЛЬНОГО ДРЕНАЖА ЛЕГОЧНЫХ ВЕН

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Врожденные пороки сердца (ВПС) составляют треть всех врожденных пороков развития и встречаются у 0,7–1,7% новорожденных детей. В структуре ВПС частота встречаемости тотального аномального дренажа легочных вен составляет 1,1–3,0%.

В данном научном обзоре проводился сравнительный анализ радиологических методов исследования, таких как эхокардиография, катетеризация полостей сердца, магнитно-резонансная томография и компьютерная томография, в диагностике и послеоперационной оценке структур тотального аномального дренажа легочных вен.

Список литературы состовляет 40 публикаций, включая в себя 1 отечественный и 39 зарубежных авторов, основанных на мета-анализе, систематических обзорах и клинических исследованиях.

Тотальный аномальный дренаж легочных вен (ТАДЛВ) обуславливает высокую смертность. Продолжительность жизни у больных с необструктивной форме ТАДЛВ выше. При необструктивной форме ТАДЛВ отмечается 50% летальность к 3 мес жизни и 80% летальность к 12 мес. Пациенты с обструктивной формой ТАДЛВ умирают в период новорожденности. У новорожденных с ТАДЛВ отмечают неблагоприятный прогноз заболевания без хирургического лечения. Только пятая часть доживает до возраста 1 года. У половины пациентов, умерших в первые 3 мес жизни, летальный исход наблюдался в первую неделю после рождения. Ранняя диагностика и точная анатомическая картина при данном виде порока играют важную роль в последующей поэтапной хирургической коррекции. ЭхоКГ в этом плане оказалась недостаточной для визуализации отдельных типов ТАДЛВ. Катетеризация сердца, в отличие от КТ, сопряжены со сравнительно высоким риском развития непосредственных и отдаленных осложнений.

ЭхоКГ является рутинным методом для первичной диагностики ТАДЛВ. КТ, в свою очередь, - «золотым стандартом» и методом выбора для адекватной пред- и послеоперационной диагностики порока. При невозможности проведения КТ, а ЭхоКГ имеет ограничения, МРТ является отличным методом выбора. Катетеризация сердца целесообразна лишь при определенных показаниях, желательно для детей старше года в виду развития серьезных осложнений.

Ключевые слова: *тотальный аномальный дренаж легочных вен, диагностика, эхокардиография, КТ, катетеризация, ионизирующее излучение.*

Түйіндеме

ӨКПЕ КӨК ТЕМЫРЛАРЫНЫҢ ТҰТАС АНОМАЛ ДРЕНАЖЫ ДИАГНОСТИКАСЫНДАҒЫ РАДИОЛОГИЯЛЫҚ ЗЕРТТЕУ ӘДІСТЕРІ

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Нәрестелерде туа біткен жүрек кемістігі барлық туа біткен кемістіктердің арасында 0,7-1,7% кездеседі. туа біткен жүрек кемістігі құрылымында өкпе көк темырларының тұтас аномал дренажы 1,1-3,0 құрайды.

Балалардың 60% өмірдің алғашқы айында жүрек ақауларынан және 25% -ы туылғанға дейін қайтыс болады.

ӨКТАД эмбриогенез кезінде сирек кездесетін даму ақауы, онда өкпе көк тамырларының бір бөлігі немесе барлығы, обструкциямен болмаса обструкциясыз, оң жақ жүрекшеге немесе жүйелік веналарға келіп жалғасады. Нәтижесінде шаршау, ентігу, өкпе артериялық гипертензиясы, цианоз және жүректің прогрессивті жеткіліксіздігі секілді белгілер пайда болады.

Бұл ғылыми шолуда жүректің жалғыз қарыншасы құрылымын диагностикалау кезіндегі эхокардиография, жүрек қуыстарының катетеризациясы, компьютерлік томография сияқты радиологиялық зерттеу әдістеріне салыстырмалы талдау жасалды.

Әдебиеттер тізіміне мета-талдау, жүйелі шолулар және клиникалық зерттеулерге негізделген 29 жарияланым, оның ішінде 1 отандық және 28 шетелдік автор кіреді.

Өкпе көк темырларының тұтас аномал дренажы жоғарғы өлімге себебші болады.

Обструкциясыз ӨКТАД бар науқастарда өмір сүру уақыты жоғарырақ болып келеді. Обструкциясыз түрінде өлім 3 айлығында 50%, ал 12 айлығында 80% құрайды. Обструкция кездескен жағдайда науқастар жаңа туған кезеңде қайтыс болады. ӨКТАД-пен туған нәресетелерде хирургиялық ем жасалмаса, қолайсыз болжам байқалады.

Тек бестен бірі ғана 1 жасқа дейін өмір сүреді. 3 айға дейінгі қаза тапқан науқастардың жартысында қайғылы жағдай туылғаннан соң бірінші аптасында байқалады. Бұл ақау кезіндегі ерте диагностика мен нақты анатомиялық көрініс кезеңді хирургиялық түзету үлкен рөл атқарады.

ӨКТАД-тың кейбір түрлері кезінде эхокардиография визуализация үшін жеткіліксіз болып келеді. Жүрек катетеризациясы КТ-ға қарағанда асқынулардың қалыптасуына тікелей тәуекел болып келеді.

Эхокардиография – бұл ӨКТАД-ты алғашқы анықтауға арналған әдеттегі әдіс, КТ – ақаулықтың операцияға дейінгі және кейінгі толық диагнозы үшін алтын стандарты болып есептеледі.

Жүректің катетеризациясы тек белгілі бір көрсеткіштер үшін және жастан жастан асқан пациенттер үшін, қарсы қайшылықтар болмаған жағдайда көрсетілген.

Түйінді сөздер: *Өкпе көк тамырларының тұтас аномал дренажы, диагностика, эхокардиография, КТ, катетеризациялау, иондаушы сәулелену.*

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Relevance:

Congenital heart disease (CHD) is a common pathology that continues to cause high mortality in children in the first year of life despite advances in cardiac surgery. 60% of children in the first month of life and 25% die from heart defects before birth [3]. Total anomalous pulmonary venous drainage (TAPVC) is a rare developmental defect during embryogenesis in which some or all of the pulmonary veins drain into the right atrium or systemic veins, with or without the obstruction of the pulmonary veins, resulting in various manifestations such as fatigue, dyspnea during exercise, pulmonary arterial hypertension, cyanosis and progressive congestive heart failure. In the structure of CHD, the frequency of occurrence, TAPVC occurs in 1.1–3.0% of infants with malformations of the cardiovascular system. With the development of surgical techniques and treatments, the results of surgical correction of TAPVC have generally improved over the past few decades. Surgical repair remains problematic, with an early mortality rate as high as 10.7%. [19]. Recently, there has been a trend towards an increase in the number of children with CHD, which is associated with an improved diagnosis of CHD [2]. In 1957, Darling and colleagues described a systematic classification for this type of anomaly [12]. In children, the frequency of TAPVC types is supracardiac - 45%; infracardiac - 25%; intracardiac - 25%; and mixed - 5% [4]. Hospital mortality in various types of TAPVC was: 14.2% for supracardiac, 11.6% for intracardial, 32.6% for infracardiac, 15.8% for mixed, and 31% for an unexplained site of the abnormal connection. The overall surgical mortality in isolated TAPVC reaches 16%. The prognosis of the course of the defect largely depends on the variant of hemodynamics. Life expectancy is higher in patients with non-obstructive TAPVC. In the non-obstructive form of TAPVC, 50% mortality is noted by three months of life and 80% mortality by 12 months. Patients with obstructive TAPVC die in the neonatal period. Newborns with TAPVC have an unfavourable prognosis of the disease without surgical treatment. Only 20% of them survive to the age of 1 year. In 50% of patients who died in the first three months, a lethal outcome was observed in the first week after birth [10]. The average life expectancy of patients with pulmonary vein stenosis is three weeks, while in patients without obstruction - an average of 2.5 months [27]. In separate studies, the age of patients with corrected TAPVC reached 48 years [13].

Materials and methods

In this review study, a comparative analysis of radiological research methods was carried out: echocardiography, catheterization of the heart cavities, magnetic resonance imaging and computed tomography in the diagnosis and evaluation of the structures of total anomalous pulmonary veins connection.

The list of literature consists of 40 publications, including one domestic and 39 foreign authors, based on meta-analysis, systematic reviews and clinical studies.

Discussion

A condition for successful hemodynamic correction and a factor that reduces the risk of an unfavourable prognosis is a sufficient size of the pulmonary arteries and preserved architectonics of the vessels of the pulmonary bed [27]. In turn, for successful correction of the defect, it is necessary to determine the exact anatomy the level of entry of the pulmonary vein collector.

There are four types of abnormal pulmonary venous outflow. The first type (type I - supracardiac) TAPVC provides outflow of the pulmonary vein through a vertical vein that flows directly into the superior vena cava. This is the most common type, affecting up to 50% of TAPVC cases. In type II (intracardiac), venous drainage flows directly into the right atrium or coronary sinus. In type III (infracardiac), the return of the pulmonary vein reaches the right atrium via the inferior vena cava. In type IV (mixed), venous connections are located both in the supracardiac and infracardiac positions [12]

The pathophysiology of TAPVC mainly depends on the degree of obstruction of the pulmonary venous flow. Infracardiac TAPVC is more likely to be obstructed due to the tortuous course of the pulmonary venous return through the liver. Patients with supracardiac TAPVC are less likely to have obstruction. Intracardiac TAPVC with the connection of the pulmonary venous return to the coronary sinus has the slightest chance of obstruction. Pulmonary venous outflow obstruction leads to severe pulmonary venous congestion, pulmonary oedema, and pulmonary arterial hypertension, leading to severe cyanosis and respiratory failure in the first few hours of life. Therefore, significant obstruction requires immediate neonatal surgery [27].

The degree of functional obstruction is also affected by the size of any interatrial communication, with less communication being associated with more severe symptoms. However, if the interatrial communication is large, it allows blood flow to the left side of the heart, reducing pulmonary hypertension. Infants with TAPVC who do not have an obstruction or have an adequate atrial septal defect may go undiagnosed in the neonatal period and appear later in childhood [2]. At present, the primary methods for studying abnormal pulmonary venous drainage

are echocardiography, multislice computed tomography, and catheterization of the heart cavities.

The advantage of echocardiography is the ability to study pathology antenatally. For example, Nandhini et al. The existence of direct and indirect ultrasound markers has been reported to raise the suspicion of antenatal TAPVC. There may be an increase in the distance between the descending aorta and the pulmonary artery in four-chamber mode. The left atrium (LA) wall may appear smooth, indicating the absence of an orifice of the pulmonary vein, or an additional vessel may be seen between the LA and the descending aorta, representing the confluence of the pulmonary veins. The right-to-left asymmetry may be more pronounced in the third trimester as pulmonary venous blood flow increases.

Ultrasound features can also help determine the type of TAPVC. The supracardiac type of TAPVC will show a dilated SVC and innominate vein. The intracardiac view of TAPVC will show an enlarged coronary sinus, while an accessory vessel seen in the sagittal section of the fetal chest and abdomen can be seen in the infracardiac type. [2].

The first line study to evaluate neonates with suspected TAPVC is transthoracic echocardiography. Transthoracic echocardiography is usually an adequate diagnostic tool to provide the surgeon with preoperative data. Oh, et al. reported that the sensitivity and specificity of transthoracic echocardiography in the diagnosis of TAPVC were 97% and 99%, respectively. In their study, the diagnosis of a particular type of TAPVC was correct in 24 out of 34 patients (71%) [28].

Transthoracic echocardiography can also evaluate the presence or absence of pulmonary venous obstruction. Doppler echocardiography can detect pulmonary vein obstruction and assess the degree of obstruction by measuring blood flow velocity in a stenotic pulmonary vein [15].

In a study of a total of 84 (83 of which were compared with surgical data) identified cases, the authors described two missed cases of PAPVR by echocardiography found after surgery to repair an atrial septal defect (ASD). Among 82 cases diagnosed as TAPVC by echocardiography, only 1 case of TAPVC was misdiagnosed as PAPVR [39].

However, Quanli Shen et al. write that transthoracic echocardiography failed to clearly show the draining veins in four children with infracardiac TAPVC and two children with mixed TAPVC due to poor acoustic windows and is less helpful in detecting distal pulmonary veins [30].

Thus, in one study of 26 people, the accuracy of echocardiographic visualization of vascular anatomy was evaluated, which could not demonstrate 4 cases of infracardiac type TAPVC. [5]

In another study, echocardiography was highly sensitive in the diagnosis of isolated TAPVC (81%), while the diagnostic value was low (27%) in the mixed variant of TAPVC associated with other CHD [15].

Thus, echocardiography may play a primary role in diagnosing cardiac anomalies, including TAPVC.

Although echocardiography remains the first-line non-invasive imaging modality for assessing congenital heart disease [34], its inherent disadvantages, such as a limited acoustic window, poor spatial resolution compared to MSCT, and operator dependence, limit its usefulness. These limitations become more pronounced in cases of postoperative sutures of the sternum and mediastinal scar tissue. Finally, echo may be insufficient when evaluating extracardiac lesions and/or vessels [35]. MSCT of the cardiovascular system can be performed at low levels of radiation exposure in patients with TAPVC. Its accuracy compared to the results of intervention studies is excellent. CT is an effective imaging modality when a non-invasive technique is desired, especially if cardiac MRI poses an increased risk to the patient [35].

Newer CT scanners provide images with better temporal and spatial resolution, greater anatomic coverage per revolution and less volume of intravascular contrast material, as well as better 2D reformation and 3D reconstruction due to the acquisition of an isotropic data set [33].

In the study by Shen et al., evaluating the role of MSCT in the diagnosis of obstructive total anomalous pulmonary venous drainage, eighteen children participated, including five with supracardiac, three intracardial, eight infracardiac and two mixed types. All of them were compared with surgical data during the operation. Thus, the diagnostic accuracy of MSCT reached 100%. [30]. In turn, the accuracy of transthoracic echocardiography was 61%. These gaps can be reduced with CT angiography. CT angiography is a good diagnostic modality for preoperative assessment of newborns and infants with TAPVC [21, 22, 25, 29]. Oh, et al. [28] compared MSCT with echocardiography in assessing TAPVC. In their study, MSCT accurately identified the location of the pulmonary vein collector drain, vertical vein stenosis, and the course of an atypical vessel into the systemic vein (sensitivity 100%, specificity 100%). The specificity of echocardiography was 100% for three findings; sensitivity, however, was 87%, 71%, and 0%, respectively.

In recent years, 3D printing technology has gained wide popularity, which significantly facilitates the understanding of the doctor's disease and the patients themselves [17]. In addition, it provides a more complete visualization of the abnormal heart. The main reason for using 3D printed models is to overcome the limitations of 2D medical imaging, which cannot fully demonstrate the spatial relationships between intracardiac structures and the geometric relationships between large vessels and surrounding anatomical structures [19]. For example, a study by Ivan Lau et al. found an excellent correlation between 3D printed models and original CT or MRI images in demonstrating normal anatomy of the heart and detecting pathologies with a mean difference of less than 0.4 mm. According to a recent systematic review, the cost associated with 3D printing varies as it depends on the materials used for 3D printing and ranges from $1 to $2,000. However, the application of this technology remains relatively limited in IHD and lacks a comprehensive overview of 3D printing in the CHD. Clinical applications of 3D printing in IHD can be divided into five main areas: preoperative planning, preoperative modelling, intraoperative orientation, medical education, and communication in medical practice. Given the data analysis from these studies, this review discusses only the three most frequently cited areas, including preoperative

planning, medical education, and model accuracy. However, time-consuming and labour-intensive image segmentation and expensive printing remain the two main limitations preventing the use of IHD 3D printing in everyday clinical practice [24].

The traditional method for diagnosing congenital heart defects is catheter angiography, which makes it possible to most accurately assess the hemodynamic and anatomical features of the pulmonary arterial and venous bed [30]. Although angiography is effective in diagnosis, it is an invasive procedure [27]. However, catheterization can lead to complications in high-risk individuals, such as arrhythmias, thrombosis, cardiac arrest, up to death [9, 23, 28, 37, 38]. Many studies prove that exposure to ionizing radiation in childhood is associated with an increased risk of developing cancer [13] since frequent and repeated cardiac catheterization procedures lead to a high cumulation of radiation doses [29, 36].

In a study by Mehta et al., 11,073 children who underwent cardiac catheterization between January 1994 and March 2006 were analyzed. In 816 studies (510 men, 63%), 858 (7.3%) complications (classified as major or minor). There were 195 major (22%) and 663 (78%) minor complications. Vascular complications were in the majority (n = 278; 32.4%) and were serious in 53 cases. Twenty-five children died within 24 hours (0.23% of the total number of cases) [32]. In turn, Bennet et al. indicate a high risk of complications in children under one year of age. Thus, in a study of 4454 cases of catheterization, the frequency of complications in infants of the first year of life was 13.9%, compared with children older than a year. Among them, 91 cases with serious complications, including four deaths.

Magnetic resonance imaging is another method to detect and describe complete or partial anomalous pulmonary artery drainage. Although transthoracic echocardiography is a first-line diagnostic tool, suboptimal acoustic windows may preclude adequate visualization of the pulmonary veins or atrial septal defect. In addition, the shortcomings of computed tomography do not allow calculation of the shunt fraction, ionizing radiation and a greater risk of nephrotoxicity when using gadolinium in cardiac MRI [8]. Anatomically, MRI provides non-invasive volumetric anatomical data and allows assessment of systemic pulmonary return and the number, origin, direction, and outflow of all pulmonary veins, including abnormal connections or obstruction. The detection rate for each pulmonary vein is 57% for the right superior pulmonary vein, 62% for the left superior pulmonary vein, 76% for the right inferior pulmonary vein, and 86% for the left inferior pulmonary vein. MRI also examines the presence and type of atrial septal defect; quantitatively determines the volume of the ventricles; evaluates the output tract of the right ventricle and the pulmonary trunk, areas of stenosis or aneurysm of the right ventricle - pulmonary arteries or branches of the pulmonary arteries; and quantifies pulmonary regurgitation and shunts, biventricular function, ejection fraction, myocardial viability, ascending aortic blood flow (including aneurysm, dissection, coarctation, evaluation of aortopulmonary collaterals and arteriovenous malformations), coronary anomalies, and coronary artery disease. Cardiac MRI is versatile for tissue characterization and has a superior ability to assess cardiovascular physiology, evaluate viability and perfusion, and detect myocardial fibrosis, as well as quantify cine and shunt fractions in complex partial anomalous pulmonary venous return compared to other imaging modalities [7].

In a study by Chang Y.C., MRI was performed on 21 patients with TAPVC. Of the 21 cases, 19 were classified as total and two as partial. Of the 19 cases of TAPVC, seven were supracardiac, nine cardiac, one infracardiac, and two mixed types. Visualization of the combined axial and coronal planes was sufficient to assess each individual pulmonary vein. The sagittal plane provided no additional information. Accurate identification of confluence of pulmonary veins and abnormal connection of pulmonary veins 95% (20/21). The limitation of echocardiography and angiocardiography makes cardiac MRI necessary for evaluating pulmonary vein-pulmonary vein fusion. This eliminates the need for invasive angiocardiography and is an essential adjunct to inadequate echocardiography [11].

As noted earlier, surgical correction of the defect is the only method that can improve the condition of patients, up to complete restoration of function. In a study by Rajamma Mathew et al. cardiac activity was assessed in 12 infants with an isolated total anomalous venous return to the lungs. Four had a severe pulmonary venous obstruction and severe pulmonary hypertension. Eight had no obvious venous obstruction and lower pulmonary pressure. In all subjects, the end-diastolic volume of the right ventricle was increased (197% of the predicted norm), and its ejection fraction was normal. The volume of the left ventricle, generally speaking, was still within the normal range. After correction, it was found that the size of the left atrium is not critical in the surgical treatment of TAPVC. Cardiac function returns to normal after surgery [31]. In the literature, there is a case of a patient 47 years old after TAPVC repair who underwent surgery for total anomalous pulmonary venous return at 14 months of age in March 1960 at Texas Children's Hospital. When this patient was voluntarily re-evaluated in 2007, echocardiography revealed long-term results of an operation that had been performed 47 years earlier: normal pulmonary vein velocity, open pulmonary venous return, and no significant electrocardiographic abnormalities. This patient is possibly the oldest known survivor of total anomalous pulmonary venous return surgery. The patient's left ventricular ejection fraction was 55% [13]. No obstruction was observed at the site of the surgical anastomosis, and the peak systolic velocity of the pulmonary veins was 54.3 cm/sec, which is within the normal range for a patient of this age [13].

Data from Jürgen Hörer et al. they say survival after 20 years is 82.7 ± 2.9%. SVH (5.9% of patients, $P<0.001$) was the only significant risk factor for mortality in multivariate analysis. Reoperations on the heart were not performed in 82.2 ± 3.3% after 20 years. Thus, there is excellent reoperation-free survival up to the third decade in patients with isolated TAPVC without the obstruction of the pulmonary veins, regardless of the type of abnormal connection. However, the survival of patients with obstruction and SVH is one of the lowest of all congenital heart diseases. Reoperation for pulmonary vein obstruction is rare and is mainly required in patients operated on in the neonatal period. Survival can be increased by using a low-flow cardiopulmonary bypass strategy [20].

Most children with TAPVC with obstruction develop symptoms in neonatal or early infancy. CT angiography is superior to transthoracic echocardiography in evaluating pulmonary venous outflow and obstruction, especially in children with infracardiac and mixed TAPVC. Cardiac catheterization is necessary only for specific indications, preferably for children older than a year. Moreover, for patients older than eight years, for the sake of radiation safety, it is desirable to have MRI without sedation or CTA. Increasingly, new technologies are being used in preoperative preparation, such as 3D printing of the heart and blood vessels.

Mortality after restoring total anomalous pulmonary venous connection has decreased but remains highest in newborns and patients with intracardiac type of connection, pulmonary vein obstruction, single ventricle of the heart. Adverse anatomical characteristics remain essential determinants of postoperative survival despite improvements in perioperative and postoperative care.

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