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TOTAL ANOMALOUS VENOUS CONNECTION

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Abstract

Introduction. Total anomalous pulmonary venous connection (TAPVC) is a rare congenital cardiac defect that occurs in less than 1% of people with congenital heart disease. It is a life-threatening defect, and without surgical treatment, few individuals survive past the first year of life. There are no known fetal environmental variables associated with TAPVC. One case series reported a 3:1 male preponderance for TAPVC to the portal vein, but not for other TAPVC locations.

Aim. In this scientific review, a comparative analysis of literature data on the epidemiology, etiology, pathogenesis of the disease, the main methods of treatment for various types of total anomalous pulmonary vein drainage was carried out.

Search strategy: Literature search was carried out in the electronic databases PubMed, MEDLINE, Web of Science, Google Scholar and e-library, using keywords (congenital heart disease, total anomalous venous connection, anatomy, morphology, embryology). Relevant data reflecting the features of this defect were taken for description in the review. Of all the selected articles for further analysis, 26 sources were included that met the inclusion criteria and excluded duplication or repetition of information.

Results. Several crucial variables have a significant impact on the etiology and clinical presentation of TAPVC. The most crucial component is the presence or absence of blockage at any level of the vein system. In blocked forms, elevated pulmonary venous pressure causes a rise in hydrostatic pressure in the capillaries, resulting in pulmonary edema. A surgical treatment is accepted as urgent if a patient also has an obstruction. Imminent fatality after surgical reconstruction is from 2% to 20%. A success between 80% and 98% can be reached after surgical treatment due to the analysis of the case series and the presence of venous obstruction

Conclusion. Knowledge of the causes of development and pathogenesis, clinic and surgical methods of treating TAPVC will allow us to detect this pathology at an early stage, to understand and improve the tactics of managing the patient, and to choose the tactics of surgical treatment correctly.

Key words: *Total anomalous pulmonary venous connection, epidemiology, embriology, types, clinical presentation, surgical treatment.*

Резюме

ТОТАЛЬНЫЙ АНОМАЛЬНЫЙ ДРЕНАЖ ЛЕГОЧНЫХ ВЕН

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Актуальность: Тотальный аномальный дренаж легочных вен (ТАДЛВ) - редкая врожденная аномалия сердца, встречающаяся только у 1% пациентов с врожденными пороками сердца. Это опасная для жизни аномалия, где лишь немногие пациенты живут дольше первого года жизни без хирургической коррекции. Факторы окружающей среды плода для ТАДЛВ неизвестны.

Цель. Провести сравнительный анализ данных литературы об эпидемиологии, этиологии, патогенезе заболевания, основных методах лечения при различных видах тотального аномального дренажа легочных вен.

Стратегия поиска:поиск литературы был осуществлен в электронных базах PubMed, MEDLINE, Web of Science, Google Scholar и e-library, по ключевым словам (врожденный порок сердца, тотальный аномальный дренаж легочных вен, морфология, эмбриология.). Релевантные данные отражающие особенности данного порока, были приняты для описания в обзоре. Из всех отобранных статей, для последующего анализа было включено 35 источников, которые отвечали критериям включения и исключали дублирование или повтор информации.

Результаты. Несколько основных факторов оказывают значительное влияние на патофизиологию и клиническую картину ТАДЛВ. Наличие или отсутствие обструкции на любом уровне венозного пути является наиболее важным фактором. При обструктивных типах высокое давление в легочных венах приводит к повышению гидростатического давления в капиллярах, что приводит к развитию отека легких. Если у пациента с ТАДЛВ также есть обструкция, это считается неотложным хирургическим вмешательством. Немедленная смертность после пластики ТАДЛВ колеблется от 2% до 20%. В зависимости от количество изученных пациентов показатель успеха лечения ТАДЛВ колеблется от 80% до 98%; успех зависит от степени обструкции легочных вен.

Заключение. Знание причин развития и патогенеза, клиники и хирургических методов лечения ТАДЛВ позволит выявлять данную патологию на ранних этапах, позволить понять и улучшить тактику ведения пациента, правильно выбрать тактику хирургического лечения.

***Ключевые слова**: тотальный аномальный дренаж, эпидемиология, эмбриология, классификация, клиника, хирургическое лечение.*

Түйіндеме

ӨКПЕ КӨК ТЕМЫРЛАРЫНЫҢ ТҰТАС АНОМАЛ ДРЕНАЖЫ

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Кіріспе: Өкпе көк темырларының тұтас аномал дренажы (ӨКТАД) — туа біткен жүрек ақауы бар пациенттердің тек 1% - ында кездесетін сирек туа біткен жүрек аномалиясы. Бұл өмірге қауіпті аномалия, онда бірнеше пациенттер хирургиялық түзетусіз өмірдің бірінші жылынан ұзақ өмір сүреді. Бірнеше негізгі факторлар

Мақсаты.Аурудың эпидемиологиясы, этиологиясы, патогенезі, жалпы аномальді өкпе веноздық дренажының түрлерін емдеудің негізгі әдістері туралы әдебиет деректеріне салыстырмалы талдау жасау.

Іздеу стратегиясы: Әдебиеттерді іздеу PubMed, MEDLINE, Web of Science, Google Scholar және e-library электронды мәліметтер базасында түйінді сөздерді (жүрек туа біткен ақауы, Өкпе көк темырларының тұтас аномал дренажы, морфология, эмбриология) қолдану арқылы жүргізілді. Шолуда сипаттау үшін осы ақаудың ерекшеліктерін көрсететін тиісті деректер алынды. Әрі қарай талдау үшін барлық таңдалған мақалалардың ішінен қосу критерийлеріне сәйкес келетін және ақпараттың қайталануын алып тастаған 35 дереккөз қосылды.

Нәтижелер. Патофизиологияға және ӨКТАД клиникалық көрінісіне айтарлықтай әсер етеді. Веноздық жолдың кез-келген деңгейінде кедергінің болуы немесе болмауы маңызды фактор болып табылады. Обструктивті түрлерде өкпе тамырларындағы жоғары қысым капиллярлардағы гидростатикалық қысымның жоғарылауына әкеледі, бұл өкпе ісінуінің дамуына әкеледі. Егер ӨКТАД -мен ауыратын науқаста да кедергі болса, бұл шұғыл хирургия болып саналады. ӨКТАД пластикасынан кейінгі жедел өлім 2% - дан 20% - ға дейін. Зерттелген пациенттердің санына байланысты ӨКТАД емдеудің сәттілік деңгейі 80% - дан 98% - ға дейін; сәттілік өкпе веналарының кедергі деңгейіне байланысты.

Қорытынды.ӨКТАД дамуының себептері мен патогенезін, клиникасы мен хирургиялық әдістерін білу бұл патологияны ерте кезеңде анықтауға, науқасты емдеу тактикасын түсінуге және жетілдіруге, хирургиялық емдеу тактикасын дұрыс таңдауға мүмкіндік береді.

***Түйінді сөздер:** Өкпе көк темырларының тұтас аномал дренажы, эпидемиология, эмбриология, жіктеу, клиника, хирургиялық емдеу.*

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Relevance.

Total anomalous venous connection.

Only 1% of people with congenital heart disease have total anomalous pulmonary venous connection, a rare congenital cardiac defect. When present, it is a fatal anomaly, and few patients survive past the first year of life without surgical repair. Significant advancements in surgical technique, cardiac anesthetic, myocardial preservation, and postoperative care for patients with this condition have been made over the previous ten years. TAPVC in newborns and young children continues to test the pediatric cardiac service's capabilities in spite of this advancement [9]. In TAPVC pulmonary veins must be connected to systemic veins and also can be combined with other cardiac abnormalities. Most of the patients died after birth before the development of the cardiac surgery system. If TAPVC is isolated there would be a significant decrease if mortality while being treated.

Aim. In this scientific review, a comparative analysis of literature data on the epidemiology, etiology, pathogenesis of the disease, the main methods of treatment for various types of total anomalous pulmonary vein drainage was carried out.

Search strategy: Literature search was carried out in the electronic databases PubMed, MEDLINE, Web of Science, Google Scholar and e-library, using keywords (congenital heart disease, total anomalous venous connection, anatomy, morphology, embryology). Relevant data reflecting the features of this defect were taken for description in the review.

As this disease is uncommon even among CHD, and because neonates are the most common patients, there have only been a few of research conducted in this field anywhere in the globe during the past ten years. The search depth had to be expanded to 21 years before we were able to gather the data we need. Of all the selected articles for further analysis, 35 sources were included that met the inclusion criteria and excluded duplication or repetition of information.

Epidemiology

TAPVC is a relatively uncommon kind of cardiac malformation, roughly seven new instances are reported for every one hundred thousand live births each year [6, 28, 29, 32]. Monogenic illnesses such as Holt-Oram and Noonan syndromes have been documented to be associated with TAPVC, despite the fact that TAPVC most commonly arises as an independent abnormality [18, 26, 32]. Despite studies of incidences of dominant inheritance in siblings and first-degree relatives [3, 16, 18, 25], the majority of patients diagnosed with TAPVC do not have a history of congenital cardiac disease in their families. There is a lack of knowledge regarding the fetal environmental variables of TAPVC. There was a frequency of TAPVC in men in the portal vein (3.6:1), but it was not seen in any other TAPVC sites, according to a case series [19]. TAPVC is associated with complex heart malformations and often contributes to the severity and poor prognosis of this disease in patients who have heterotaxia syndrome. Heterotaxia syndrome is a syndrome of malformations that is characterized by abnormal left-/right-sidedness of the chest and abdomen. On the other hand, the vast majority of cases of TAPVC are not linked to any other important heart abnormalities than foramen ovale and patent ductus arteriosus.

Embriology.

During embryonic development, one of the organs that begins differentiating and functioning the earliest is the heart. The heart originates from the cardiogenic mesoderm. Vascular development can take place in a number of locations, but it is particularly obvious when the heart is first taking shape [10, 26, 33]. The fast expansion of the heart is what gives the early embryo its characteristic "bulge" appearance. As it forms inside the germinal disc, in the beginning stages it appears as a simple couple of vessels within the newly developing pericardial cavity. By folding this disc, the heart can assume an anatomically correct position in the chest [10, 34]. The umbilical system, the system of the developing fetus, and the yolk system are the three systems of the aortic and venous circulation that are distinguishable in the blood vessels of the embryo. The extraembryonic mesoderm of the chorion is the main in the placental system development. It's responsible for eradicating debris produced by the foetus and acts as a conduit for nutrient delivery to the mama. This channel stop its function after baby birth. Both of the embryo come the point where the embryonic system starts, which will transfigure into a completely formed circulatory system. The vitelline system, which lines the outer layer of the yolk sac and facilitates blood flow between the gut and the liver, is found in all multicellular organisms [10, 34]. The lungs originate as protrusions from the foregut. Later they divide from the plexus. During this period the lungs are not connected to the heart [24, 29, 32, 34]. At the 25–27 days of pregnancy, the venous plexus of the lungs keeps links to the right superior vena cava, the left superior vena cava, and the portal system. These links are maintained for the remainder of the pregnancy. Right and left atriums do not connect to each other. During gravid days 27 to 29, the major pulmonary vein protrudes from the left atrium. Ultimately, this growth will make a connection to the major pulmonary venous system. The right superior vena cava and azygos vein grows form the right common cardiac system. The left superior vena cava and the coronary sinus develop after they enter to the left venous sinus from the left common cardiac vein. The rise to the inferior vena cava, ductus venosus, and portal vein are given from this system [10, 32]. The pulmonary venous system forms by the joining of the primary pulmonary vein and the pulmonary venous plexus join by the end of the first month of pregnancy. The posterior wall of the left atrium is supplied with blood by the common pulmonary vein. Other connections resolve over time in a healthy person [10, 16, 29, 34].

TAPVC forming an aberrant connection leads to the absence or incomplete formation of a connection between the common pulmonary vein and the venous system of the lungs. This leads to expansion of the right atrium and right ventricle, due to pulmonary venous return [29]. The continuation of the right and left cardinal veins as well as the umbilical vein are all factors that lead to the development of atypical links between the veins of the lungs [2]. Mortality is more likely to take place during the month after birth if the lungs do not have access to a pulmonary or a systemic link throughout that time [2]. An atrial septal defect or an open foramen ovale is an essential component for life support, as blood enters the left ventricle through them. Also, the pathology may include an open ductus

arteriosus, as one of the components [22, 23]. Among TAPVC cases, 75% have a extensively rank communicating tone without venous return blockage [2]. Again, 25% of individualities witness confined and dammed systemic venous rotation [2]. TAPVC may coexist with other cardiac anomalies such as transposition of the great arteries, with or without discordance, left superior vena cava accessory etc. Also, asplenia can be seen in patients with TAPVC in ¾ cases [23]. In the Baltimore Washington Infant Study from 1981 to 1987, 522 newborns with CHD were analyzed which 1.5% had TAPVC, 1/5 of cases were with extracardiac pathology [5, 14].

Types.

TAPVC Without PVO

Symptoms are often mild at birth, and cyanosis may be mild if there is a free departure of pulmonary blood flow through the aberrant pulmonary junction back into the right atrium and subsequently through the foramen ovale. All of the blood from the pulmonary and systemic circulations mingle together in the right atrium, which results in a full admixture. Massive pulmonary overcirculation arises in the first few days of life as a result of a reduction in the pulmonary vascular resistance that occurs during the first few days of life. The increased volume and pressure are then passed to the right atrium and the right ventricle. By redirecting blood from the right atrium to the left atrium, cardiac output may be kept stable. The severity of cyanosis varies, but the oxygen saturation can get up to the middle of the 90s, which can be explained by a pulmonary-systemic shunt that is more than 5:1. Tachypnea and feeding difficulties emerge after delivery, followed by substantial developmental delay and, in most instances, mortality before the age of one year. Although the symptoms are modest at birth, they progress as follows: Patients may appear with symptoms like hepatomegaly, tachycardia, and tachypnea. During the cardiac examination, there is either no murmur or the presence of a quiet pulmonary ejection murmur in addition to the typical presence of an S3 gallop. An elevation can be seen on the electrocardiogram in the right atrium and ventricle. Additionally, the image of the right leg of the His bundle can occasionally be seen as being incomplete. The process of echocardiography is outlined in [16, 20].

TAPVC With PVO

Significant changes are brought about in the clinical picture, physiology, and prognosis as a result of the existence of PVO, which can be found in anywhere from 25 to 50 percent of patients who have TAPVC [12, 13, 16, 17]. The level of obstruction can range from a moderate elevation in pulmonary venous pressure all the way up to pulmonary venous atresia, which is associated with life-threatening respiratory failure. In the first few hours after birth, pulmonary venous hypertension and edema progress in unison with the rise in pulmonary blood flow. These changes often take place within the first few hours following delivery. Pulmonary edema causes reflex vasoconstriction of the pulmonary arteries in addition to reduced gas exchange, all of which contribute to progressive hypoxemia in the long run. It is not always easy to tell the symptoms apart from those of newborn respiratory distress syndrome, and the results of chest x-rays might be similar at times. In the initial few hours after delivery, obstructed TAPVC may present with only mild symptoms; nevertheless, the condition has a tendency to worsen with time, leading to a progressive course of hypoxemia and acidosis in spite of intensive therapy. In circumstances in which there is extensive TAPVC obstruction, the degree of hypoxia can range from mild to severe, but it is virtually always severe and does not react well to further oxygenation. This is because severe hypoxia does not respond well to additional oxygenation. The physiologic signs of obstructive TAPVC often entail severe respiratory failure but more modest cardiac abnormalities [8]. On the other hand, tachycardia and hepatomegaly are symptoms that nearly never fail to appear in this illness. There is a possibility that the precordium has a murmur.

There are four types of this type of congenital heart desease:

Supracardiac type, the abnormal connection occurs at the supracardiac level, and the left innominate vein is commonly involved. Other uncommon supracardiac venous connections include the superior vena cava (SVC) and azygos veins (AZs).

Cardiac type, when pulmonary veins connected to the right atrium or to the coronary sinus.

Infracardiac type refers to an improper connection into the portal veins, ductus venous, hepatic veins, or inferior vena cava.

In the mixed type at least two distinct regions are drained by the pulmonary veins.

In their announcement, Herlong and colleagues presented a categorization scheme with increased granularity. This system has a crucial connection to the morphological and physiological changes that occur in TAPVC. These alterations involve the parameters listed below: (1) the degree of connections, which may be classified as supracardiac, cardiac, infracardiac, or mixed; (2) the presence or absence of a blockage; and (3) the origin of the obstruction, which can be classified as extrinsic, intrinsic, or obstructive atrial septal communication [4, 11, 16, 35].

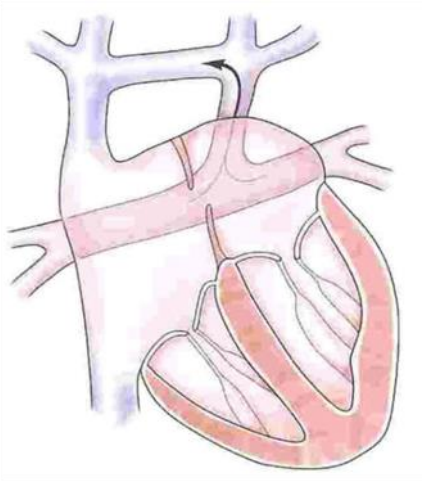

Figure 1. Supracardiac TAPVC: The common pulmonary vein drains superiorly into the innominate vein or superior vena cava via an ascending vertical vein. Bove EL, Hirsch JC. Total anomalous pulmonary venous drainage and cor triatriatum. In: Gardner TJ, Spray TL, editors. Operative Cardiac Surgery, London: Arnold Publishers; 2004:581–592.

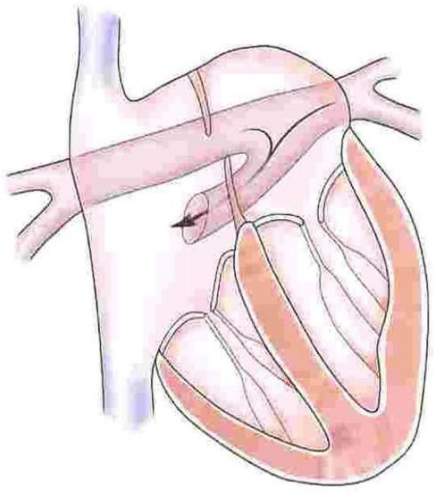

Figure 2. Cardiac TAPVC: The common pulmonary vein drains into the coronary sinus or, on rare occasions, individual pulmonary veins will connect directly into the right atrium. Bove EL, Hirsch JC. Total anomalous pulmonary venous drainage and cor triatriatum. In: Gardner TJ, Spray TL, editors. Operative Cardiac Surgery, London: Arnold Publishers; 2004:581–592.

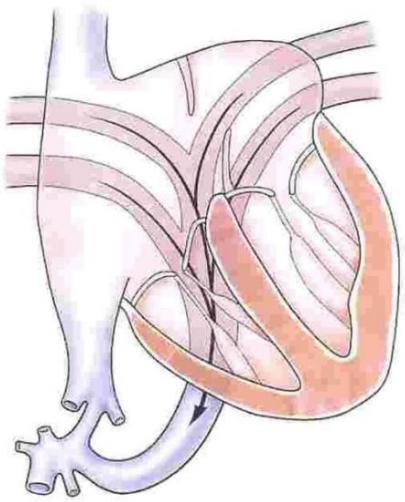

Figure 3. Infracardiac TAPVC: The common pulmonary vein drains through the diaphragm into the portal vein or ductus venosus. Bove EL, Hirsch JC. Total anomalous pulmonary venous drainage and cor triatriatum. In: Gardner TJ, Spray TL, editors. Operative Cardiac Surgery, London: Arnold Publishers; 2004:581–592.

Pathophysiology.

It is the mixing of oxygen-rich blood from the pulmonary system with deoxygenated blood from the systemic venous circulation that is the primary cause of the hemodynamic alterations that take place during TAPVC. Cyanosis and a lack of oxygen are the results of this in babies. Since this is the case, TAPVC is the sixth most prevalent cause of cyanotic heart disease.

The pathogenesis of TAPVC, as well as its clinical appearance, is significantly influenced by a number of important external variables. The presence or absence of blockage at any level of the venous route is the factor that is considered to be the most critical. There are multiple locations where obstructions can take place, including: (1) a confluent vein that is traveling through tissue that causes external compression similar to intrathoracic structures (supracardial type) or the diaphragmatic inlet (infracardial type); (2) internal compression that is the result of a narrowing of the lumen; and (3) the location where confluent blood enters the systemic venous return pathway.

The intracardiac form is nearly often linked with blockage, which typically takes place as a result of a confluent vein making a vertical entry via the esophagus into the diaphragm of the patient's chest. It's not very often that the intracardiac kind is accompanied with blockage. However, there is still a possibility of finding a blockage at the CS or at the entry to the RA. There is an association between venous blockage and one-half of the supracardiac instances of TAPVC.

In addition to this, obstructive TAPVC can be caused by a narrowing of the lumen of the left innominate vein, the SVC, or the azygos vein. Additionally, the passage of a vertical vein between the left pulmonary artery and the left bronchus, which results in external compression [27], is a probable cause of blockage. This is one of the possible causes of obstruction.

The extent of the patient's interatrial communication is a key factor to consider when evaluating individuals who have non-obstructive TAPVC. After delivery, pulmonary resistance begins to fall, and a suitable amount of blood begins to reach the pulmonary bed to facilitate healthy oxygen exchange. In the right atrium, there is a combination of blood that has been saturated and desaturated. When a newborn has a condition known as nonrestrictive atrial communication, also known as a big atrial septal defect (ASD), blood is able to enter the left ventricle of the heart and supply the infant's systemic circulation. Even though blood is being redirected from the right to the left side of the body, there is still 3-5 times more blood entering the pulmonary bed, and the pressure in the pulmonary artery steadily increases. In neonates, hypercirculation ultimately results in right ventricular hypertrophy, right ventricular failure, and desaturation.

When it comes to obstructive kinds, a high pressure in the pulmonary veins causes a rise in the hydrostatic pressure in the capillaries, which in turn causes the development of pulmonary edema. At the same time, increasing pressure in the pulmonary artery causes inadequate blood flow in the pulmonary capillaries. When there is no quick relief coming from the clogged artery, severe desaturation might develop.

TAPVC can present itself in a broad variety of ways, and these symptoms are very variable depending on the degree of blockage and the pulmonary vascular resistance. Within the first 12 hours of a newborn's life, acute sickness characterized by tachypnea, tachycardia, dyspnoea, hypoxemia, and metabolic acidosis develops when there is a significant blockage present. If the surgical repair cannot be made, death might occur during the first few days of the condition. On the opposite end of the range, patients who do not have venous blockage are often asymptomatic at birth, but they may experience tachypnea, moderate cyanosis, and difficulty feeding in the first few weeks of their lives. There is a progressive onset of profound developmental delay as well as repeated infections of the respiratory system, and only a tiny percentage of patients

are able to survive until late childhood or adolescent if they do not receive therapy [7].

Clinical presentation. TAPVC Without Pulmonary Venous Obstruction

Patients could not exhibit any symptoms when they are first born. In most cases, the symptoms of hypercirculation in the lungs will become apparent during the first few days of a person's existence. Babies may have difficulty breathing, problems with eating, and a delay in their developmental progress. The severity of cyanosis varies from patient to patient, and some people may have just a minor form that is asymptomatic. Patients will, at some point in the future, develop hypertrophy of the right ventricle as well as symptoms of right ventricular failure. At the time of the physical examination, the patient was found to have cyanosis, a fixed split S2, and an ejection systolic murmur. There is a possibility that signs of right ventricular hypertrophy and/or insufficiency are present, such as elevation of the right ventricle, a loud P2, hepatomegaly, and the presence of a third heart sound [1].

TAPVC with Pulmonary Venous Obstruction

The newborn phase is typically marked by acute respiratory failure and cyanosis in patients diagnosed with obstructive TAPVC. The presence of hypoxia, hypotension, and tachypnea are all possible results from a physical examination. This is a common alternative diagnosis that is considered to be a possibility in cases of chronic pulmonary hypertension in infants. These infants are typically ill and require emergency surgery as soon as possible. TAPVC is most often found to exist as a standalone heart lesion, however it can also be seen in conjunction with other congenital heart disorders. Patients who have heterotaxy with polysplenia or asplenia have a greater risk of developing TAPVC, according to studies [1, 15].

Surgical treatment

Still, it's considered a surgical exigency, if a case who has TAPVC also has an inhibition. Immediate mortality after TAPVC form ranges from 2% to 20%. Different sources give different information about the success rate, which ranges from 80% to 98%, depending on the form of TAPVC [2, 22, 23]. In 5-10% of cases, a fatal outcome is observed due to obstruction of the pulmonary veins, despite this, success can reach 90% [31]. The postoperative course is affected by the anesthetic agents used, quantum of time on cardiopulmonary bypass (CPB), aorticcross-clamp time, depth of hypothermia, and duration of circulatory arrest [21]. Bypass alone affects coagulation, platelet function, electrolyte balance, glycemic control, and extravascular fluid accumulations, which affects the length of time the case will have to remain in the neonatal ferocious care unit (NICU) [21]. In an imperative situation, the immediate surgical ideal is to connect the common pulmonary venous channel to the left atrium, divide the perpendicular pulmonary tone, and closeinter-arterial shunts, if present. The surgery demands much of a perioperative nanny in terms of association, planning, and prosecution. The cscrub person, and anesthesia care provider insure that input and affair are rigorously covered throughout the intraoperative phase. In all situations, not just extremities, the arterial conduit is linked and twice ligated incontinently after CPB is established. Theinter-arterial shunt must be ligated before form to help air from entering the systemic rotation, which could beget cerebral damage [31]. Both a single venous and single arterial cannula are used for all forms of TAPVC. The following are the general way for surgical form of the main types of TAPVC (ie, supracardiac, cardiac, infracardiac).

SUPRACARDIAC TAPVC: The surgeon ligates the left-sided vertical vein's connection to the innominate vein. Access is gained to the left atrium, and the confluence of the pulmonary veins in the left atrium is anastomosed for the venous fusion (Figure 4). Concurrently, the ASD and exposed foramen ovale are closed. After ligating the venous confluence, the surgeon performs a direct connection with the posterior part of the left atrium [30, 31].

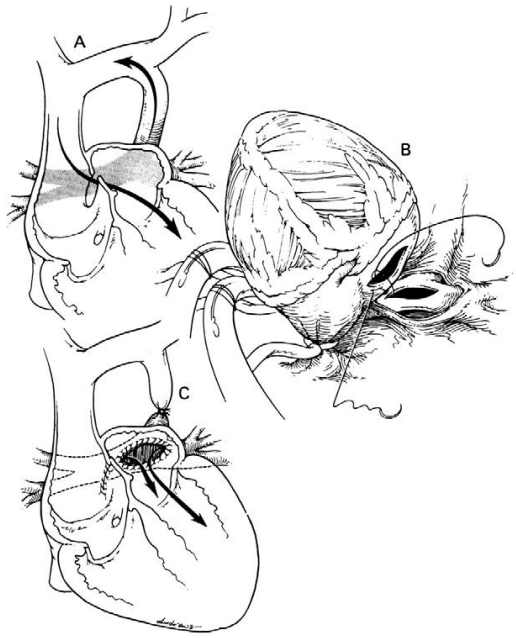

Figure 4. Steps in the repair of supracardiac total anomalous pulmonary venous connection.

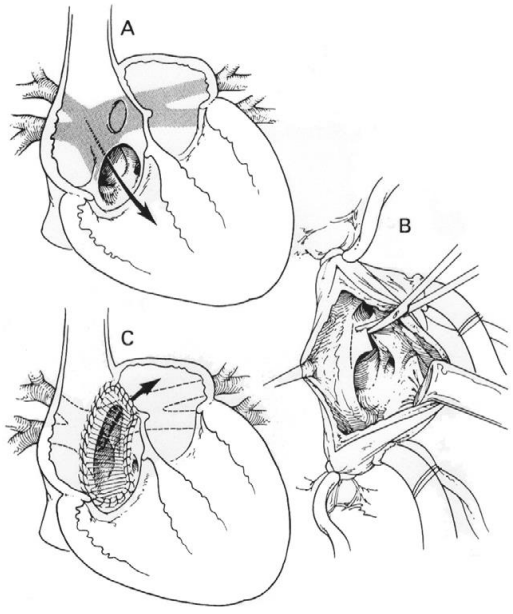

Figure 5. Steps in the repair of a total anomalous pulmonary venous connection to the coronary sinus.

CARDIAC CONNECTION TO THE CORONARY SINUS: The surgeon makes space for a large atrial junction by slicing the coronary sinus septum and the primary septum. When separating the left and right atria, a patch is used to seal the newly formed ASD and the entrance of the coronary sinus. The absence of the coronary sinus permits all pulmonary veins and the coronary sinus to return to the left atrium [30, 31] (Figure 5).

CARDIAC CONNECTION TO THE RIGHT ATRIUM: The surgeon removes a portion of the atrial septum and attaches a patch to the defect's borders and the right atrium's posterior wall. This bypasses the defect by rerouting blood from the pulmonary veins directly into the left atrium (Figure 6).

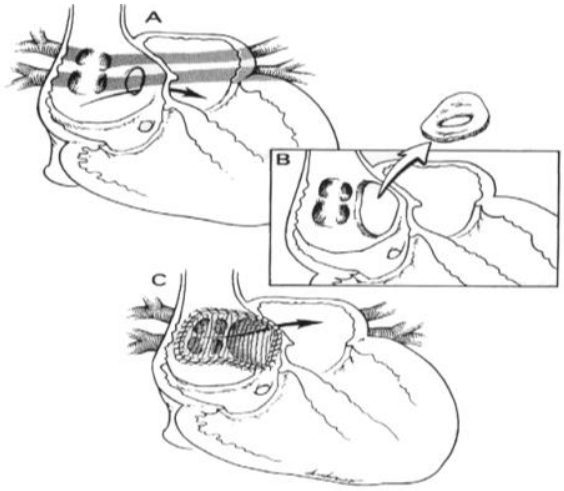

Figure 6. Steps in the repair of cardiac total anomalous pulmonary venous connection to the right atrium.
(Figures 4, 5, and 6 from Reardon M.J., Cooley D.A., Kubrusly L. et al. Total anomalous pulmonary venous return: report of 201 patients treated surgically. *Tex Heart Inst J.* 1985; 12:131-141).

INFRACARDIAC CONNECTION: After establishing cardiopulmonary bypass, the surgeon ligates the descending vertical vein where it enters the diaphragmatic hiatus to avoid future flow of blood. To get to the left atrium, an incision is made in the heart's posterior wall. Additionally, the surgeon creates an anastomosis between the left atrium and the venous confluence by making a cut in the anterior wall of the venous confluence. Following the incision, the patent ductus is subsequently sealed (Figure 7).

Conclusion. Total anomalous pulmonary venous connection is a rare but life-threatening congenital heart disease. The study of the causes of development and pathogenesis, clinics of this pathology will allow to identify this pathology at an early stage, to understand and improve the tactics of patient management. Also, with various types of defect, it makes sense to use various surgical methods for the treatment of TAPVC to correctly choose the tactics of surgical treatment.

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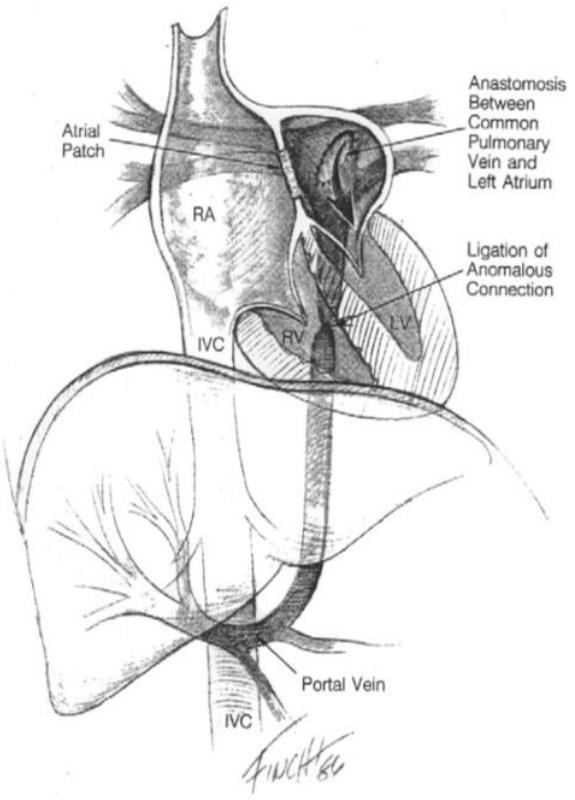

Figure 7.Surgical correction of infradiaphragmatic total anomalous pulmonary venous connection. RA = right atrium; RV=right ventricle; LV = left ventricle; IVC = inferior vena cava
(From Amplatz K., Moller J.H. The Radiology of Congenital Heart Disease. St Louis, Mo: Mosby Year Book; 1993).

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