

دهمین همایش

قلب کودکان ایران

10th Iranian Congress of Pediatric Cardiology

08-10 February 2024, Tehran



با امتیاز
بازآموزی

۱۹-۲۱ بهمن ۱۴۰۲

هتل قلب شهید رجایی

08 - 10 February 2024

Rajaie Heart Institute - TEHRAN, IRAN



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Syncope in the Pediatric Population: Evaluation and Outcomes

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Syncope is defined as a transient loss of consciousness resulting from temporary global cerebral hypoperfusion, characterized by rapid onset, short duration, and spontaneous complete recovery. It can be classified into three main categories: neurally mediated (reflex syncope), orthostatic hypotension, and cardiac syncope.

When approaching the diagnostic evaluation of syncope, it is crucial to differentiate it from other causes of loss of consciousness and establish the underlying diagnosis. This involves conducting a detailed patient history, performing a physical examination, obtaining a standard 12-lead electrocardiography (ECG), and measuring orthostatic blood pressure.

The treatment of syncope focuses on addressing the underlying cause. The overall morbidity and mortality associated with syncope are generally low, except in cases of cardiac syncope. Patient reassurance plays a vital role in the treatment process.

For neurally mediated syncope, reassurance and patient education are often sufficient interventions. Physical therapy and the use of drugs may be considered in resistant cases, and in some instances, a permanent pacemaker (PPM) may be necessary.

In cases of orthostatic hypotension, management strategies include gradually transitioning from a supine or sitting position to standing, increasing fluid and sodium intake, and discontinuing or reducing the dosage of medications that contribute to the condition.

Cardiac syncope may require the use of antiarrhythmic drugs, cardiac pacing, catheter-directed ablation, or, in rare cases, an implantable cardioverter-defibrillator (ICD).

In conclusion, an organized approach to the diagnostic evaluation and treatment of syncope is crucial for managing this condition effectively. Proper identification of the underlying cause and implementation of appropriate interventions can greatly improve patient outcomes.

بیماری های مزمن
مزمن

Chronic Diseases

اکو جینین کاردیوگرافی
اکو جینین

Fetal Echocardiography

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Common Cardiac Channelopathies: Management and Challenges

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Long QT Syndrome (LQTS) is a hereditary arrhythmogenic disorder characterized by abnormally prolonged QT intervals on electrocardiograms (ECGs), which can lead to life-threatening ventricular arrhythmias, including torsades de pointes. This abstract provides a summary of key aspects of LQTS, including its prevalence, genetic basis, clinical features, diagnostic criteria, and management strategies.

LQTS affects approximately 1 in 2000 live births, making it a relatively rare but clinically significant condition. Genetic studies have identified at least 13 genes associated with LQTS. Among these genes, mutations in three major genes—KCNQ1 (LQT1), KCNH2 (LQT2), and SCN5A (LQT3)—account for approximately 75% of cases with a strong clinical phenotype.

LQTS can present with various symptoms. Some individuals may have prolonged QT intervals without experiencing any symptoms (asymptomatic). However, others may experience palpitations, irregular heartbeats, or fluttering sensations. Fainting episodes (presyncope and syncope) can also occur due to arrhythmias, and in severe cases, life-threatening arrhythmias can lead to cardiac arrest. In rare cases, LQTS may be associated with neuronal deafness (Jervell and Lange-Nielsen syndrome) or skeletal defects, such as those seen in Andersen-Tawil syndrome or Timothy syndrome.

Several challenge tests are employed to evaluate and assess the susceptibility to arrhythmias in individuals with LQTS. These tests include:

- **Brisk Standing ECG Changes:** An orthostatic challenge that provokes arrhythmias.
- **Epinephrine Infusion:** Used during stress tests to assess susceptibility to arrhythmias.
- **Exercise Test:** Evaluates the response of the QT interval during physical exertion.

In conclusion, understanding the prevalence, genetic associations, clinical features, and challenge tests in LQTS is crucial for diagnosing and managing this potentially life-threatening cardiac channelopathy.

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Approach to Wolff-Parkinson-White (WPW) Syndrome

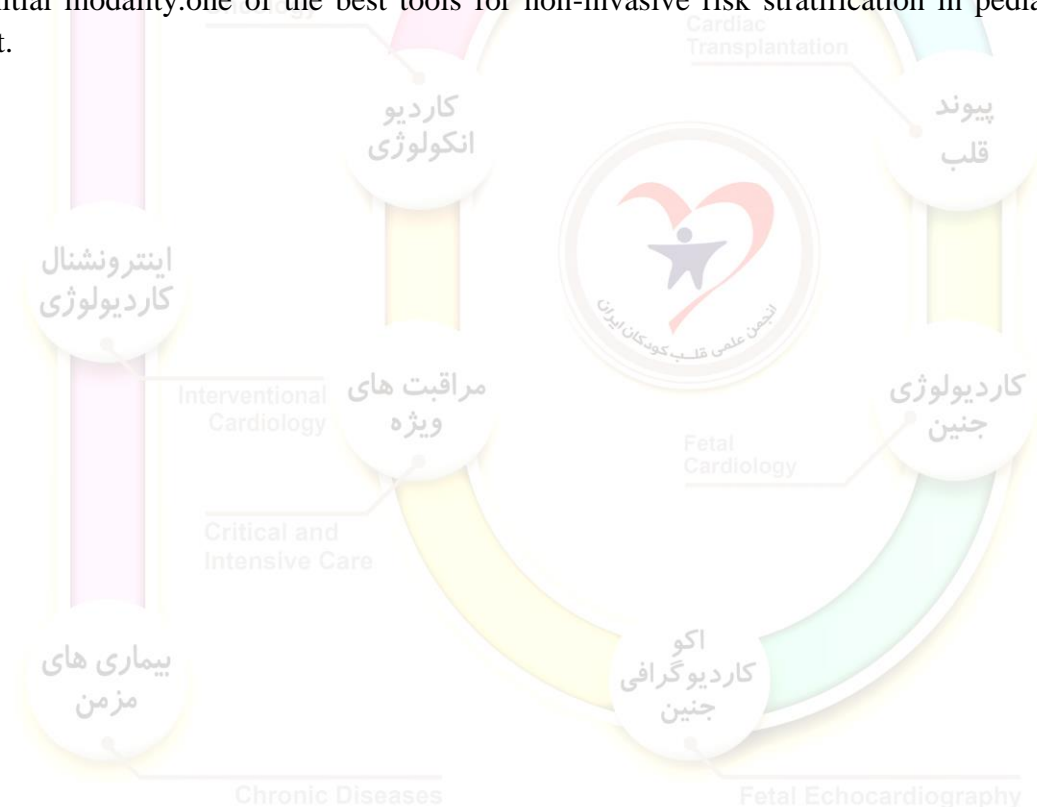
Mohammadrafie Khorgami MD

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Wolff Parkinson White Syndrome (WPW) is a congenital abnormality that involves the presence of abnormal electrically conductive circuits between the atria and ventricles. The disorder includes accessory electrical pathways that bypass the AV node.

the general prevalence of WPW has been estimated between 1 to 3 per 1000 individuals (0.1 to 0.3 %).

studies have been shown the rate of sudden cardiac death between 0.0002 to 0.0015 per patient-years for patients with WPW pattern. High-risk features on clinical evaluation include male sex, WPW pattern detected in the first two decades of life or less than 35 years, history of atrial fibrillation (AF), arrhythmic symptoms (especially syncope), congenital heart disease (e.g., Ebstein's anomaly), or familial WPW syndrome. Risk stratification of asymptomatic WPW pattern may be performed either invasively or by non-invasive means. Neither risk-stratification scheme is 100% perfect. Non-invasive evaluation is usually a preferred initial modality. one of the best tools for non-invasive risk stratification in pediatric group is exercise test.



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Electrophysiological Interventions in Structural Heart Anomalies

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Introduction:

Cardiac electrophysiologists frequently encounter cases where traditional approaches are inadequate due to anatomical variations or congenital abnormalities. In such cases, a comprehensive understanding of the cardiac anatomy and its conduction system, knowledge about previous interventions, and access to specialized procedure facilities are essential.

Challenges in Ablation Procedures in Abnormal Cardiac Anatomies:

Ablation procedures in abnormal cardiac anatomies pose several challenges. These include difficulties in catheter placement and maneuverability, limitations in electrical signal interpretation and localization during mapping, strategies for lesion creation despite anatomical obstacles, and the need to safeguard adjacent critical structures during ablation.

Epstein's Anomaly of the Tricuspid Valve and Mapping and Ablation of Accessory Pathways (APs):

In Epstein's anomaly, the most frequent locations of APs are the posterior septal, posterolateral, and right free wall. Precise localization of the atrioventricular (AV) groove can be achieved through RCA angiography, and the use of long steerable sheaths is required for catheter stabilizing. Ablation is performed at the atrial side of the accessory pathways at the AV groove.

Abnormal Cardiac Positions:

Dextrocardia encompasses various sub-types and is rarely a mirror image of a normally oriented heart. Ablation procedures in patients with dextrocardia pose challenges due to abnormal access, atrioventricular connections, chamber arrangements, ventricular-atrial positions, and arterial locations. Although the conduction system in dextrocardia patients is consistently abnormal, its anatomy can be understood and predicted with the aid of advanced imaging tools.

Ablation in Univentricular Hearts:

Univentricular hearts exhibit significant diversity in cardiac anatomies. Accurate determination of the configuration of the cardiac conduction system is crucial in these cases. Identifying the precise mechanisms of arrhythmias can be challenging and time-consuming.

Conclusion:

Successful ablation of cardiac arrhythmias in patients with abnormal anatomy requires a comprehensive understanding of anatomical variations and tailored approaches to overcome associated challenges. By utilizing advanced imaging techniques, promoting multidisciplinary collaboration, and employing specialized tools, improved outcomes and patient care can be achieved.

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Ductal Stenting in Newborns with Less Than 2 kg of Weight

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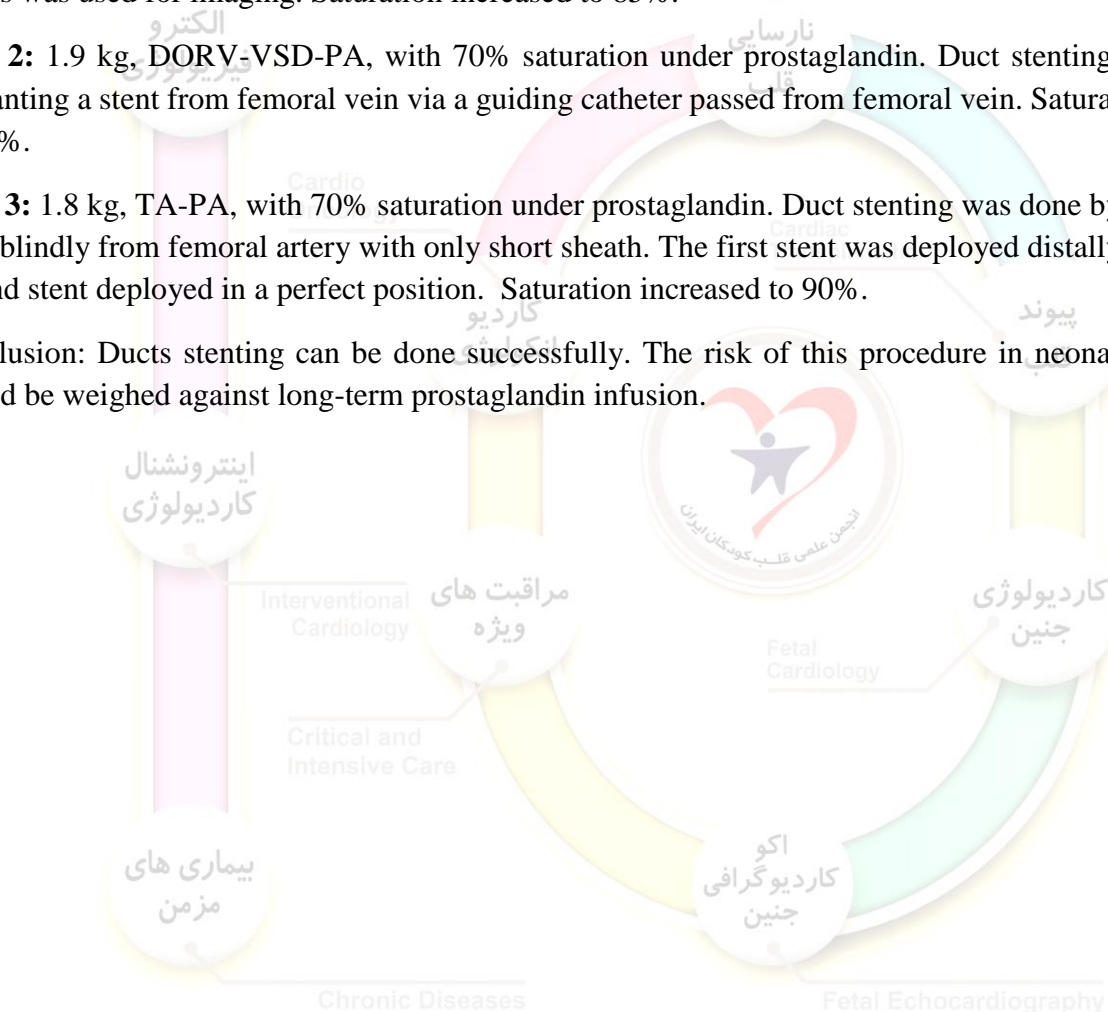
Introduction: Ductal stenting is the preferred palliative procedure for neonates with duct-dependent pulmonary circulation. In neonates with weight under 2 kg, this procedure is not easy, so in developed countries long term prostaglandin infusion and waiting weight gain is recommended. However, this strategy is not feasible in our country. So we developed methods for duct stenting in these patients and so far performed in 3 patients.

Case 1: 1.9 kg, DORV-VSD-PA, with 62% saturation under prostaglandin. Duct stenting was done by implanting a stent from femoral artery via only short sheath. A NIH catheter passed from femoral vein access was used for imaging. Saturation increased to 85%.

Case 2: 1.9 kg, DORV-VSD-PA, with 70% saturation under prostaglandin. Duct stenting was done by implanting a stent from femoral vein via a guiding catheter passed from femoral vein. Saturation increased to 92%.

Case 3: 1.8 kg, TA-PA, with 70% saturation under prostaglandin. Duct stenting was done by implanting a stent blindly from femoral artery with only short sheath. The first stent was deployed distally in RPA. The second stent deployed in a perfect position. Saturation increased to 90%.

Conclusion: Ducts stenting can be done successfully. The risk of this procedure in neonates under 2kg should be weighed against long-term prostaglandin infusion.



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Hypoplastic Aortic Arch Stenting in Neonates

Mohammadreza Edraki M.D.

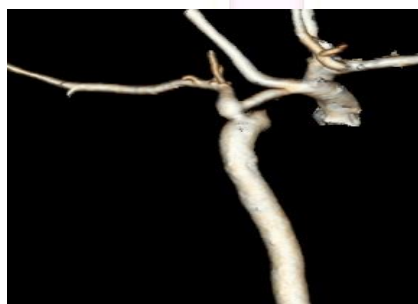
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Background: Management of hypoplastic aortic arch in newborns and infants is controversial(1). The stenosis might present with cardiogenic shock, renal dysfunction, and metabolic acidosis, and can deteriorate promptly (2).

Surgical repair is treatment of choice, unless the case is ill with unstable hemodynamic, although the operation of hypoplastic arch may impose hazardous effects particularly for brain (3).

Method: One of our cases was a 20 days old boy, 2800 gram, who presented with pulmonary edema and metabolic acidosis. Echocardiography and CT angiography showed double outlet right ventricle (with large ventricular septal defect physiology) and hypoplastic arch (Fig 1), whose condition was not suitable for surgical correction.

Fig 1: CT angiography of the patient before angiography:



Thus we decided to insert a formula stent 535 6*25 into the stenotic segment, but the minimal delivery system French size for insertion of the device was 6 and it was too big to insert from the femoral artery.

So we proceeded retrogradely from IVC to the right ventricle and then VSD toward the aortic arch (Fig 2-A), and opened the stent just after brachiocephalic artery, while covered the both left common carotid and subclavian arteries. The stent dilated the arch successfully, while the subclavian artery became stenotic (Fig 2-B). Hence we steered and opened a non-compliant coronary balloon 3*15 through the stent zigs (Fig 2-C).

Results: The outcome was acceptable (Fig 2-D), with 5 mm Hg pressure gradient across the arch, and 6 months after the procedure, he was in desirable condition with 15 mm Hg pressure gradient.

Conclusion: Aortic arch stenting might be applicable among neonates in critical situations.

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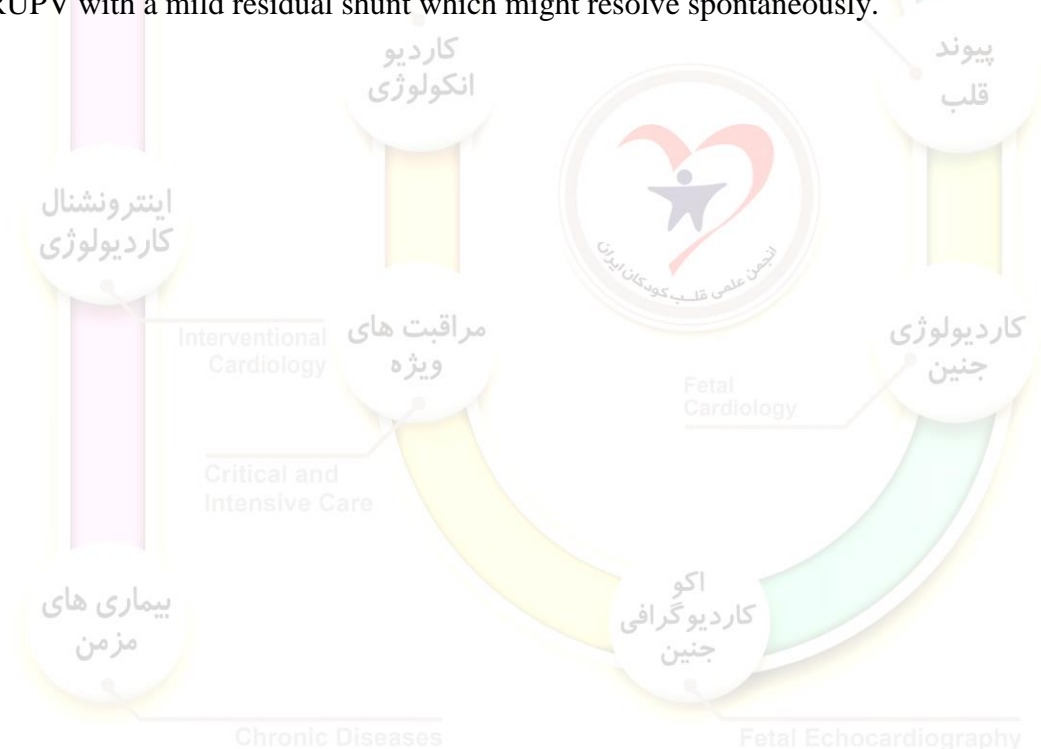
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Transcatheter Closure of Sinus Venosus ASD

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A sinus venous atrial septal defect (SVASD) is a type of congenital heart defect that predominantly involves an opening between the Superior Vena Cava (SVC) and the Right Upper Pulmonary Vein (RUPV). Surgical closure of SVASD has been the standard treatment; however, the surgical closure of SVASD is far more complex than mere ASD secundum reconstruction, and complications are more likely. Transcatheter closure of SVASD is an emerging alternative for surgical repair. We report five cases of successful transcatheter closure of SVASD and one case of failure that required surgical intervention. All patients underwent pre-procedure Computed Tomography Angiography (CTA) to determine the size and location of the defect and the optimal stent size and position. The SVC stenting was performed using balloon-expandable stents, followed by RUPV angioplasty if needed. Final angiograms and pressure measurements in the SVC, RUPV, and Right Atrium (RA) confirmed the absence of residual shunt and pulmonary venous obstruction. One patient experienced stent migration to the pulmonary artery, which necessitated surgical retrieval and defect closure. The balloon expansion test is not mandatory before stent implantation, as in the case of RUPV obstruction, its flow could be reestablished by ballooning and/or stent implantation within the RUPV with a mild residual shunt which might resolve spontaneously.



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Restrictive Cardiomyopathy and Its Differential Diagnoses

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Background: Restrictive cardiomyopathy (RCM) is a rare form of heart muscle disease characterized by impaired ventricular filling and reduced diastolic volume. Despite normal or near-normal systolic function and wall thickness, RCM poses significant challenges in diagnosis and management.

Methods: We present an overview of RCM, focusing on its definition, epidemiology, etiology, and pathophysiology.

Definition: The World Health Organization (WHO) defines RCM as restrictive filling and reduced diastolic volume of either or both ventricles. The most recent classification emphasizes the phenotypic appearance as morpho-functional, highlighting abnormal compliance without another predominant phenotype of right ventricular (RV) or left ventricular (LV) dilatation, hypertrophy, or systolic dysfunction.

Epidemiology: RCM accounts for only 2.5% to 5% of all cardiomyopathies. The average age at diagnosis is 6 years. The condition affects both males and females equally. Positive family history is observed in 25% to 30% of cases, while sporadic occurrences are also reported. Genetic factors play a crucial role, including sarcomeric protein gene defects and nonsarcomeric protein gene defects.

Etiology: Endomyocardial fibrosis is the predominant cause of RCM in children, particularly in tropical regions. Idiopathic cases are common in non-tropical areas. Amyloidosis is the primary cause in adults, although it is rarely seen in pediatric cases. Myocarditis can manifest solely with diastolic dysfunction. Diabetic children should be routinely evaluated for diastolic function. Patients undergoing radiotherapy or chemotherapy, such as anthracycline, should also be assessed for diastolic function, irrespective of systolic function.

Pathophysiology: In RCM, there is limited or no filling during late diastole (atrial systole). Ventricular compliance is decreased. Small changes in volume lead to marked ventricular pressure rise. Dysfunction or delay in active relaxation of the ventricle (involving energy-requiring processes for calcium uptake into the sarcoplasmic reticulum) contributes to the pathophysiology, rather than intrinsic stiffness of the ventricular wall.

Conclusion: A comprehensive understanding of RCM is essential for early diagnosis, appropriate management, and improved outcomes. Further research is needed to explore novel therapeutic approaches and enhance patient care.

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Fetal Cardiac Function

Fariba Rashidi Ghader MD

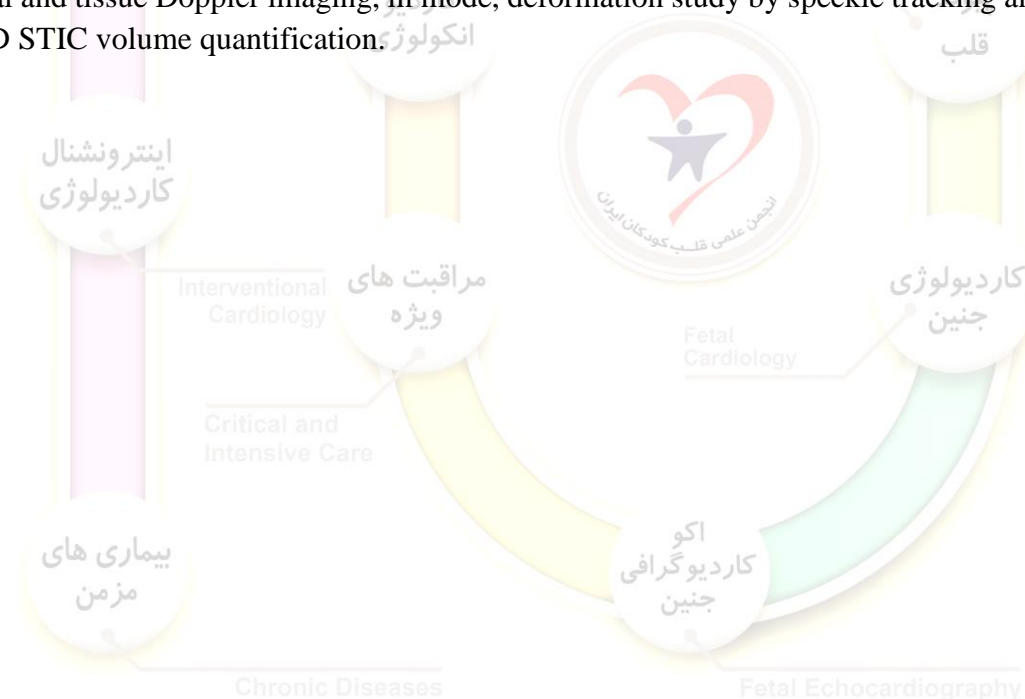
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Heart failure is inability of the heart to supply sufficient blood flow to meet the body's needs. Utility of determining fetal cardiac function is for differentiating various types of CMPs and predicting perinatal mortality in CHD. It is also a marker of disease severity and used for monitoring and predicting outcome in some diseases like IUGR, TTTS, hydrops, fetal arrhythmia, GDM or CDH and for optimizing timing of delivery. We also need to evaluate fetal cardiac function for determining necessity of intervention.

Cardiac dysfunction in fetus classified as , a) High output lesions like anemia, TTTS, vascular tumors, b) Increased preload such as complete AVSD, Ebstein, TOF with absent pulmonic valve, c) Increased afterload like aortic or pulmonary stenosis, D) External compression which leads to decreased preload such as intrathoracic SOL & pelvic mass.

All kinds of abovementioned fetal conditions cause different cardiac findings in echocardiography such as cardiomegaly, AV valve regurgitation, diastolic dysfunction and later systolic dysfunction and also can cause increased plasticity index of DV and UV notching and hydrops.

Echocardiography has been helped to assess fetal cardiac function by various techniques included 2 D, conventional and tissue Doppler imaging, m mode, deformation study by speckle tracking and color coded TDI and 4 D STIC volume quantification.



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Prenatal Diagnosis of Coarctation of the Aorta and Interrupted Aortic Arch

Maryam Moradian MD

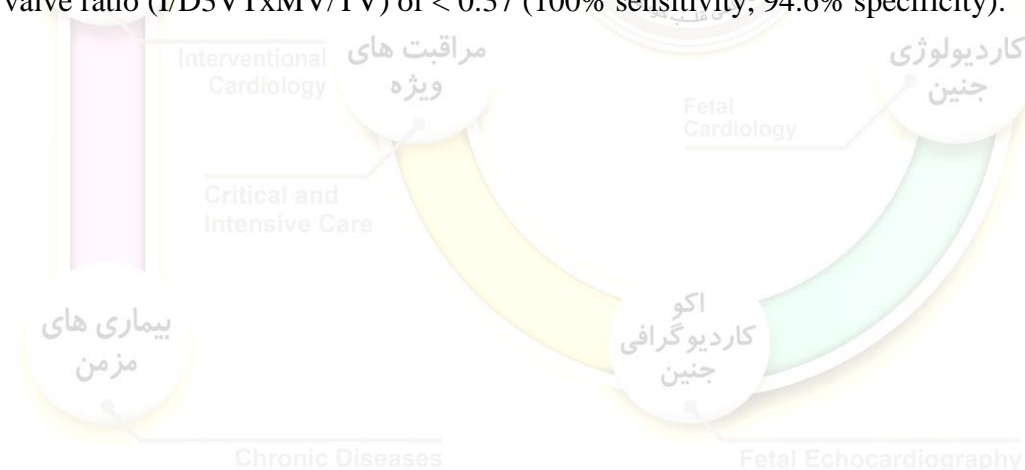
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Coarctation of the aorta (CoA) is one of the most challenging forms of congenital heart disease to diagnose prenatally. Prenatal detection can improve survival by facilitating early institution of treatment and timely surgical repair. Unfortunately, coarctation of the aorta may go undetected even after birth and during the neonatal period.

Many subjective echocardiographic markers for the prediction of coarctation of the aorta during fetal life have poor specificity, including disproportion in the size of the ventricles and great vessels, hypoplasia of the transverse aortic arch/isthmus, presence of a coarctation shelf, isthmic flow disturbance, persistence of the left superior vena cava, presence of a bicuspid aortic valve.

These subjective findings have contributed to the high false positive and false negative rate associated with antenatal detection of CoA. In particular, the assessment of cardiac asymmetry in the third trimester is difficult, as mild asymmetry can be a normal physiological response to hemodynamic changes.

Objective ultrasound assessment with multiple measurements may have higher sensitivity and specificity when compared to subjective assessment in the prenatal diagnosis of CoA. MV Z-score, TV Z-score, LV/RV ratio, width and length ratios, MV/TV ratio, AV Z-score, PV Z-score, AV/PV ratio, Aoasc/DAo ratios, I/D ratio, and Carotid-subclavian artery index (CSAI) (aortic arch diameter at the left subclavian artery, to the distance between the left carotid artery and the left subclavian artery). The most sensitive and specific predictors are: A carotid-subclavian artery index (CSAI) of < 0.78 (92.3% sensitivity, 96.8% specificity) and A product of isthmus-to-duct ratio in the three-vessel trachea view (3VT) and the mitral-to-tricuspid valve ratio ($I/D_{3VT} \times MV/TV$) of < 0.37 (100% sensitivity, 94.6% specificity).



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Fetal Arrhythmia: Differential Diagnosis and Management

Mohammad Dalili MD

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Introduction:

Fetal arrhythmias, diagnosed in approximately 1-3% of pregnancies, account for a significant proportion of referrals to fetal cardiology services. Understanding the type and mechanism of the arrhythmia is essential for appropriate management and prognosis.

Diagnostic Tools:

Fetal electrocardiogram (ECG) signal recording is limited in availability, leading to the widespread use of echocardiography for evaluating fetal atrial and ventricular activity. Magnetocardiography (MCG), a noninvasive technology utilizing electromagnetic signals from the fetal heart, is a promising tool with increasing accessibility.

Magnetocardiography (MCG):

Fetal MCG (fMCG) records the natural electromagnetic signal produced by the fetal heart. Recent advancements in sensor technology and magnetic shields have made fMCG more cost-effective, facilitating broader adoption. Similar to postnatal ECG, fMCG allows for precise assessment of cardiac time intervals and provides insights into signal characteristics and rhythm patterns.

Echocardiography for Evaluation:

Echocardiography plays a crucial role in evaluating fetal heart rate, rhythm, and atrioventricular (AV) conduction. Doppler waves and M-mode pictures provide valuable information for diagnosis and management.

Main Steps for Management:

Proper management of fetal arrhythmias involves ensuring high-quality recordings of atrial and ventricular activation signals, making a precise diagnosis of the underlying mechanism, and providing appropriate therapeutic recommendations based on the diagnosis.

Conclusion:

Accurate diagnosis is paramount in the management of fetal arrhythmias. Once the diagnosis is established, there are ample data and guidelines available to guide informed decision-making for optimal management strategies.

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Perinatal Care in Critical Fetal Heart Diseases

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Critical congenital heart disease is defined as a congenital heart condition needing surgery intervention or leading to this within 1 month after birth.

1-Historically, CHD lesions with ductal-dependent systemic or pulmonary circulations were classified as “critical” CHDs.

There is now a shift to recognize critical CHD lesions as those in prenatally which a PGE1 infusion is insufficient for hemodynamic stabilization.

2-disease-specific resuscitation in the delivery room and rapid transport for emergent cardiac intervention in the first few hours after birth:

Obstructed TAPVC- HLHS /IAS -D-TGA with restrictive atrial septum

Severe Ebstein anomaly, low cardiac output, and circular shunt - TOF/APV

Uncontrolled tachyarrhythmias or CHB with hydrops

Fetal Cardiac Catheter Intervention:

Critical aortic stenosis - pulmonary atresia with intact ventricular septum

balloon atrial septostomy for HLHS or restrictive inter-atrial communication.

Deterioration after starting PGE1:

1-pulmonary venous or left atrial obstruction

2-These include obstructive (usually infra diaphragmatic) TAPVC

3-a restrictive atrial septum HLHS, cor-triatratrium, severe mitral stenosis

4- D-TGA associated with restrictive atrial shunting

5-These patients require urgent echocardiography,

6-followed by interventional cardiac catheterization or surgery

Antenatal Detection of Treatable Critical Congenital Heart Disease is Associated with Lower Morbidity and Mortality

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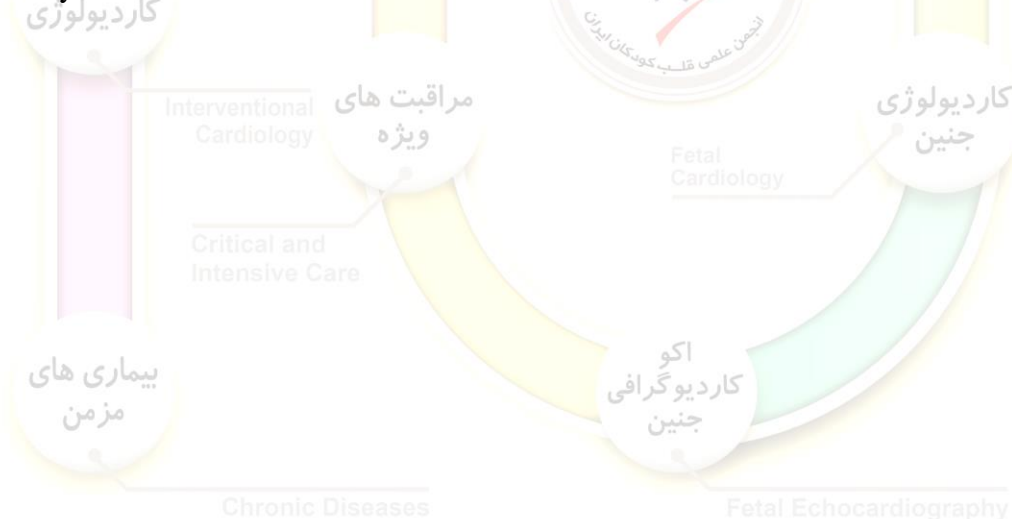
Early Postoperative management in complicated Glenn shunt

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In bidirectional Glenn anastomosis or cavopulmonary connection, the superior vena cava (SVC) connects to the ipsilateral pulmonary artery branch. If bilateral SVCs exist, the anastomosis occurs on both sides. This procedure, performed with or without cardiopulmonary bypass, addresses intracardiac issues such as atrial septectomy, atrioventricular valve regurgitation, and subaortic stenosis. Intracardiac manipulation requires aortic cross clamping, particularly for aortic arch repair. Patients with Glenn shunt may face complications like a subtle rise in pulmonary vascular resistance during the postoperative period in the intensive care unit. Impediments to pulmonary blood flow can result from elevated PVR, PA branch stenosis, myocardial dysfunction, AV regurgitation, or stenosis of Glenn anastomosis. Desaturation immediately after Glenn shunt surgery can also be caused by hypovolemia and anemia.

To determine the cause of complications in Glenn shunt cases, healthcare professionals utilize measurements of transpulmonary pressure gradient, PA pressure, and echocardiography assessment. Additionally, laboratory examinations and ECG monitoring are part of their intensive care unit (ICU) protocols. Early postoperative desaturation in Glenn shunt patients may result from hyperventilation, constricting cerebral arterioles. Maintaining PaCO₂ in the 40s mmHg range is crucial to optimize cerebral and pulmonary blood flow. Following the Glenn procedure, elevated SVC pressure can alter cerebral artery autoregulation, leading to symptoms like headaches and irritability in young infants. Adjustment to this condition typically takes 48 to 72 hours and can be eased with the administration of narcotics and NSAIDs.



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How to Achieve a “Good Fontan”, Return to the “10 Commandants” The Cardinal Role of Pulmonary Vasodilators

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Single ventricle patients are a group of heterogenous complex cyanotic congenital heart diseases which are one of the most challenging groups of CHD regarding the management. Historically many valuable efforts have been performed in this way, from Dr. William Glenn to Dr. Francis Fontan, Dr. Guillermo O. Kreutzer, and the others, till now.

The patients after single ventricle operations (Glenn and Fontan) have a new specific blood circulation in which by eliminating the sub-pulmonary ventricle, the blood enters directly from the SVC and IVC to the pulmonary vascular bed, then continues to the LA, single V and aorta: the “Fontan circuit”. Impairment of this circuit causes the single ventricle patients to face many challenges, including complications in some organ/systems such as: cardiovascular, pulmonary, lymphatic, GI/liver, renal, endocrine and neurologic systems. It seems that the basic underlying mechanisms of such the impairment are elevated CVP, lymphatic congestion and low cardiac output.

Currently, the best management for preventing these complications are not well studied and established while there are limited options for the routine medical management of Fontan patients. In this way, for constructing a systematic approach for management of single ventricle patients after Glenn/Fontan operations, we can use the “Alain Choussat 10 Commandants” in a new way, as a “hemodynamic backbone” for optimizing the “Fontan circuit”. If we consider the components of “Alain Choussat 10 Commandants”, we can target the sites of the medical/procedural/surgical intervention for optimizing the “circuit” and so achieving good clinical situation. It seems that among all components of the “Fontan circuit”, the PAP/PVR, as the “gate of the circuit”, is a critical determinant of perfect working of the Fontan hemodynamics.

According to this critical importance of PAP/PVR in Fontan circuit, it seems that pulmonary vasodilators can have a cardinal hemodynamic role in perfect working of the circuit, and so optimizing the clinical condition of the patients. Even in the patients with suitable PAP before Fontan (< 15 mmHg), some evidences revealed that there is significantly high risk for increasing PAP after the procedure, because of some factors; so we can have a hemodynamical explanation for using pulmonary vasodilators in all single ventricle patients, before Glenn shunt, interstage and after Fontan operation. The evidences regarding such the use of pulmonary vasodilators in all single ventricle patients (including the patients with suitable PAP) are very spare, limited to some evidences about the non-suitable PAP.

Based on the above mentioned hemodynamical mechanism, some evidences, and our experiences, we recommend the use of pulmonary vasodilators, from the two main groups; phosphodiesterase inhibitors: Sildenafil/Tadalafil and endothelin receptor inhibitors: Bosentan/Macitentan, before the Glenn shunt, interstage and after Fontan operation. For the patient with suitable PAP Sildenafil/Tadalafil can be used with low to medium dose, considering the condition of other components of the “Fontan circuit”, and in the patients with non-suitable PAP, combination of medium to high dose of Sildenafil/Tadalafil with Bosentan/Macitentan will be effective in achieving a good clinical situation.

Finally, we can have a systematic approach for medical management of Fontan patients, according to the components of “Fontan circuit”. The pulmonary vasodilators have a cardinal role in medical management of single ventricle patients, also the drugs such as ASA/Warfarin/anti X, diuretics, ACE inhibitors, digoxin and the others, have also important roles.

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Post Cardiac Surgery Coagulopathy

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Cardiopulmonary bypass (CPB) induces activation of the hemostatic and coagulation systems, which can lead to life-threatening bleeding. The etiology of CPB-related coagulopathy is multifactorial and complex, involving several interrelated factors:

1. **Hemodilution from CPB Circuit Prime:** Dilution of blood components due to priming of the CPB circuit.
2. **Whole Blood Loss from Surgical Bleeding:** Surgical procedures during CPB can result in significant blood loss.
3. **Tissue Factor Release:** Surgical trauma and exposure to air release tissue factor into circulation.
4. **Contact Activation of the Hemostatic System:** Interaction with foreign surfaces activates clotting pathways.
5. **Hyperfibrinolysis from CPB:** Excessive breakdown of fibrin clots.
6. **Reduced Platelet Count and Function:** Platelet dysfunction due to CPB.
7. **Inflammation:** Inflammatory responses may exacerbate coagulation abnormalities.

To manage microvascular bleeding and reverse coagulation abnormalities, recent guidelines recommend the following measures:

1. **Maintain Normothermia:** Reduce heat loss to achieve and sustain normal body temperature.
2. **Optimal Tranexamic Acid Dosing:** Balance efficacy and safety of this antifibrinolytic agent.
3. **Balanced Ratios of Blood Components:** Administer red blood cells, fresh frozen plasma (FFP), and platelets in appropriate proportions.
4. **Use of Fibrinogen Concentrate:** Consider it as a first-line treatment option alongside Prothrombin Complex Concentrate.

For cases of uncontrollable microvascular bleeding that do not respond to conventional therapy and local hemostatic materials, the following strategies can be employed:

- **Recombinant Activated Factor VII**
- **Factor VIII Inhibitor Bypassing Activity Complex:** Contains clotting factors II, IX, X, and activated factor VII.
- **Concentrates of Clotting Factor XIII**
- **Tamponade or “Packing” of Large Wound Areas**

This review comprehensively discusses the etiology and management of coagulation disorders following CPB.

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Mechanical Ventilation Following Cardiac Surgery in Children

Ahmad Jamei Khosroshahi MD

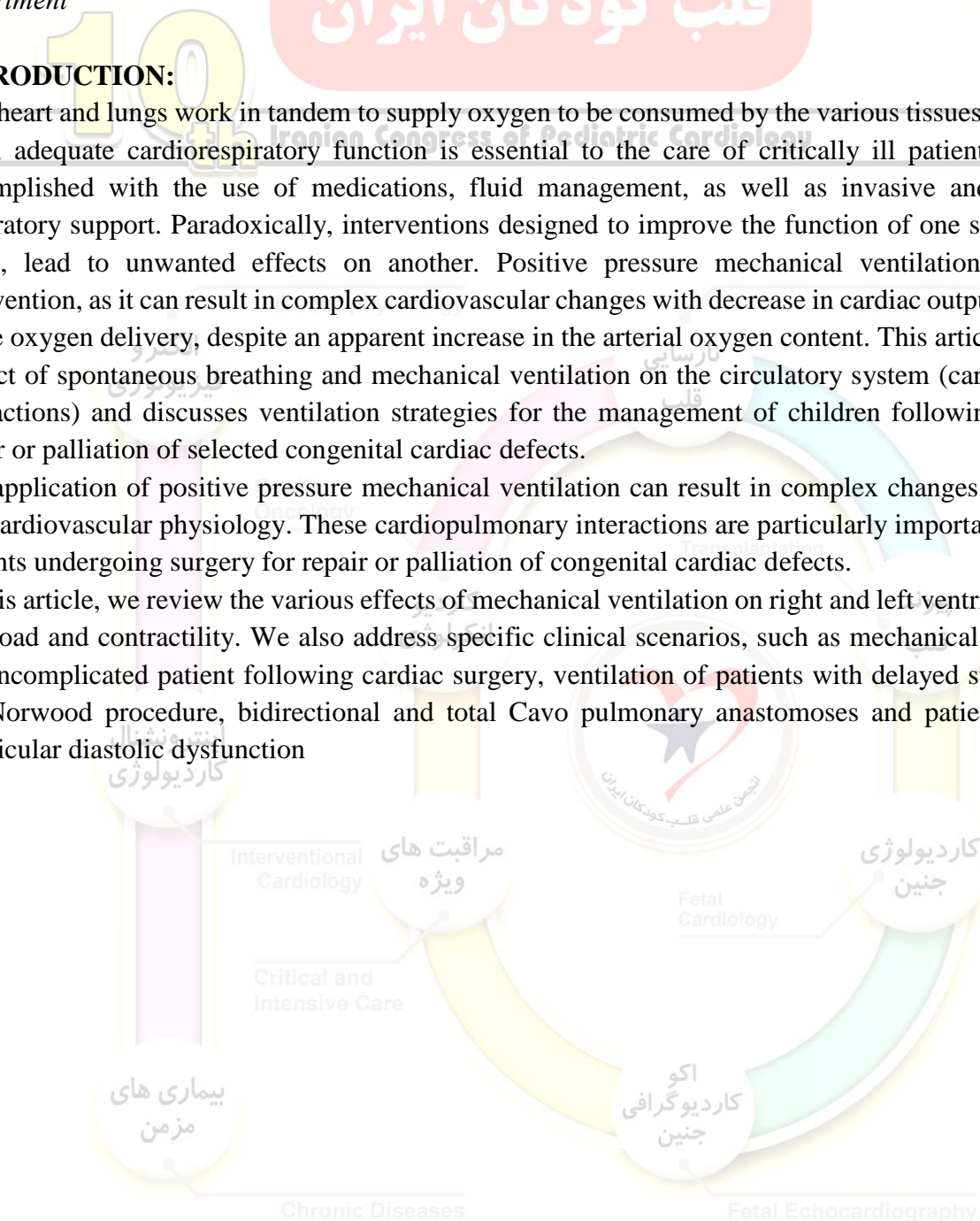
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INTRODUCTION:

The heart and lungs work in tandem to supply oxygen to be consumed by the various tissues. Maintenance of an adequate cardiorespiratory function is essential to the care of critically ill patients and can be accomplished with the use of medications, fluid management, as well as invasive and noninvasive respiratory support. Paradoxically, interventions designed to improve the function of one system may, at times, lead to unwanted effects on another. Positive pressure mechanical ventilation is one such intervention, as it can result in complex cardiovascular changes with decrease in cardiac output and reduced tissue oxygen delivery, despite an apparent increase in the arterial oxygen content. This article reviews the impact of spontaneous breathing and mechanical ventilation on the circulatory system (cardiorespiratory interactions) and discusses ventilation strategies for the management of children following surgery for repair or palliation of selected congenital cardiac defects.

The application of positive pressure mechanical ventilation can result in complex changes in pulmonary and cardiovascular physiology. These cardiopulmonary interactions are particularly important in pediatric patients undergoing surgery for repair or palliation of congenital cardiac defects.

In this article, we review the various effects of mechanical ventilation on right and left ventricular preload, afterload and contractility. We also address specific clinical scenarios, such as mechanical ventilation of the uncomplicated patient following cardiac surgery, ventilation of patients with delayed sternal closure, the Norwood procedure, bidirectional and total Cavo pulmonary anastomoses and patients with right ventricular diastolic dysfunction



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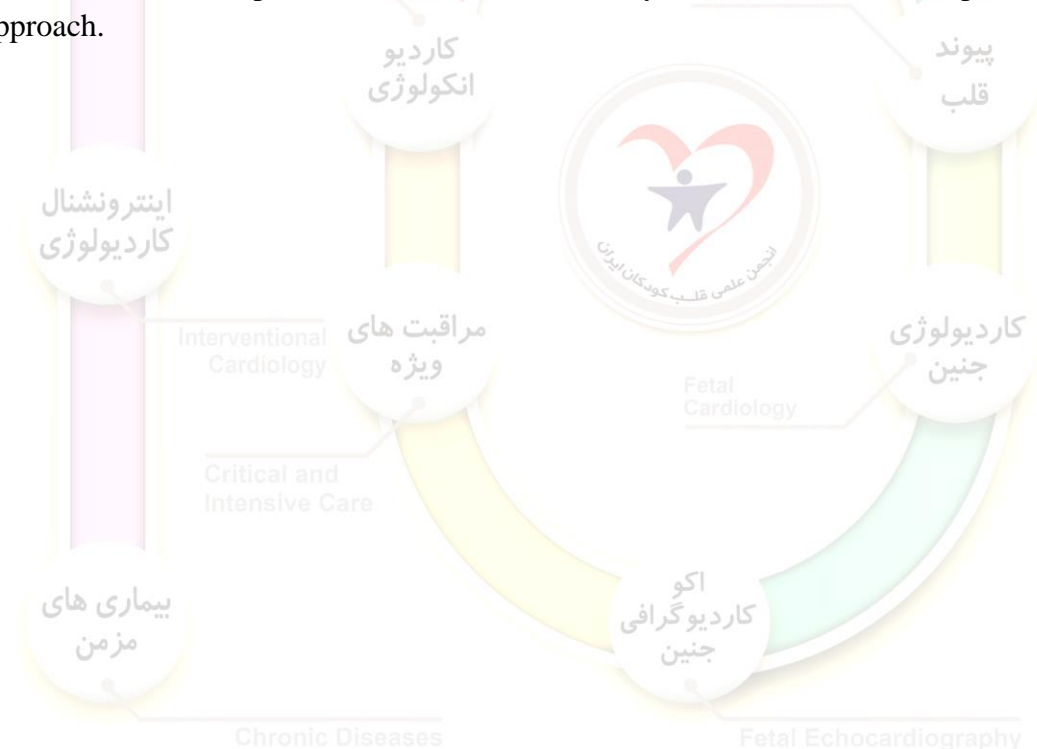
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Indications and updates in Device Closure of Septal Defects

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Transcatheter device closure for septal defects has emerged as a less invasive alternative to surgical repair. Transcatheter device closure represents a promising strategy for managing various septal defects, offering improved outcomes and reduced invasiveness. However, each defect type presents unique challenges, and ongoing research will enhance our ability to address a broader spectrum of defects using this approach. This approach offers several advantages, including lower complication rates, shorter hospital stays, and reduced mortality compared to surgical repair. Complex ASDs, such as those with large defects, rim deficiencies, fenestrated defects, multiple defects, or associated PAH, can pose challenges for device closure. Transcatheter device closure has also become an accepted alternative for some types of ventricular septal defects (VSDs). However, limitations remain, and further innovations are needed to expand the range of defects that can be safely closed via a transcatheter approach. Transcatheter device closure represents a promising strategy for managing septal defects, offering improved outcomes and reduced invasiveness. Continued research and development will enhance our ability to address a broader spectrum of defects using this approach.



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Off-Label Applications of Occluder Devices

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Abstract: Considering the size and shape of occluder devices and their associated delivery systems, these tools can be utilized in off-label scenarios. In this article, we introduce several specific applications:

1. **Membranous VSD Amplatzer for Aortopulmonary Window Closure:** The Membranous VSD Amplatzer device, with a waist length of 3 mm, features discs on both sides that are 4 to 8 mm larger than the waist. This design allows for a greater distance from the coronary arteries. Consequently, it is a more suitable option for closing aortopulmonary windows compared to the Amplatzer devices used for patent ductus arteriosus (PDA) and ventricular septal defect (VSD) closure.
2. **PiccoloADO II & Amplatzer for Coronary Fistula Closure:** Both the PiccoloADO II and Amplatzer devices are well-suited for coronary fistula closure due to their low-profile delivery systems, device flexibility, shape adaptability, and a range of available sizes.
3. **Muscular VSD Amplatzer & ASD Amplatzer for Main Pulmonary Artery Closure in Single-Ventricle Patients Post Glenn or Fontan Procedures:** In patients with single ventricles and either pulmonary stenosis (PS) or pulmonary atresia (PA band), the bulky design of the Muscular VSD Amplatzer and ASD Amplatzer devices ensures proper placement within the narrowed pulmonary artery. The presence of discs on both sides allows for secure positioning before release.

This review provides detailed insights into the etiology and management of coagulation disorders following cardiopulmonary bypass (CPB).



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Updates on Mitral Valve Prolapse

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Abstract: Mitral valve prolapse (MVP) is the most common valvular abnormality, affecting approximately 2-3% of the population in the United States. While MVP typically follows a benign course, it can occasionally lead to serious complications, including clinically significant mitral regurgitation (MR), infective endocarditis, and sudden cardiac death.

Key findings on echocardiography include:

1. **Classic MVP:** Parasternal long-axis view reveals greater than 2 mm of superior displacement of the mitral leaflets into the left atrium during systole. Leaflet thickness is at least 5 mm.
2. **Non-Classic MVP:** Displacement exceeds 2 mm, but the maximal leaflet thickness remains less than 5 mm.

MVP can be further categorized based on its underlying pathology:

- **Myxomatous MVP (Barlow's Disease):** Characterized by excess tissue, including chordal thickening/elongation, annular dilation, and calcification. Chordal rupture is less likely.
- **Fibroelastic Deficiency (FED):** Characterized by chordal thinning, elongation, and/or a high probability of rupture. FED is the most common form of MVP.

The arrhythmic mitral valve complex is defined by the presence of MVP combined with frequent and/or complex ventricular arrhythmias, including: $\geq 5\%$ total premature ventricular contraction (PVC) burden
Complex arrhythmias (non-sustained ventricular tachycardia, sustained ventricular tachycardia, or ventricular fibrillation)

Risk stratification for sudden cardiac death (SCD) and ventricular arrhythmias includes:

- **Unexplained Syncope**
- **Severe MR**
- **Left Ventricular Ejection Fraction (LVEF) < 50%**
- **Bi-leaflet myxomatous MVP**

This comprehensive review provides insights into the etiology, echocardiographic findings, and risk assessment for patients with MVP. We summarize evidence and guidelines on targeted therapies of ventricular arrhythmias in the setting of MVP, including implantable cardioverter defibrillators and catheter ablation. Our review highlights current knowledge on diagnosis, prognostic impact, and optimal management of patients with arrhythmic MVP.

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Kawasaki Disease: Understanding and Management

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Kawasaki disease (KD), also known as mucocutaneous lymph node syndrome, is a medium vessel vasculitis primarily affecting the coronary arteries. Here's a comprehensive overview and management guidelines for this condition:

Epidemiology: More common in children younger than five, with a slight male predominance. Siblings of affected individuals have a higher risk.

Pathogenesis: Triggered by an infectious agent, likely respiratory, leading to immune activation, cytokine release, and vascular lesions.

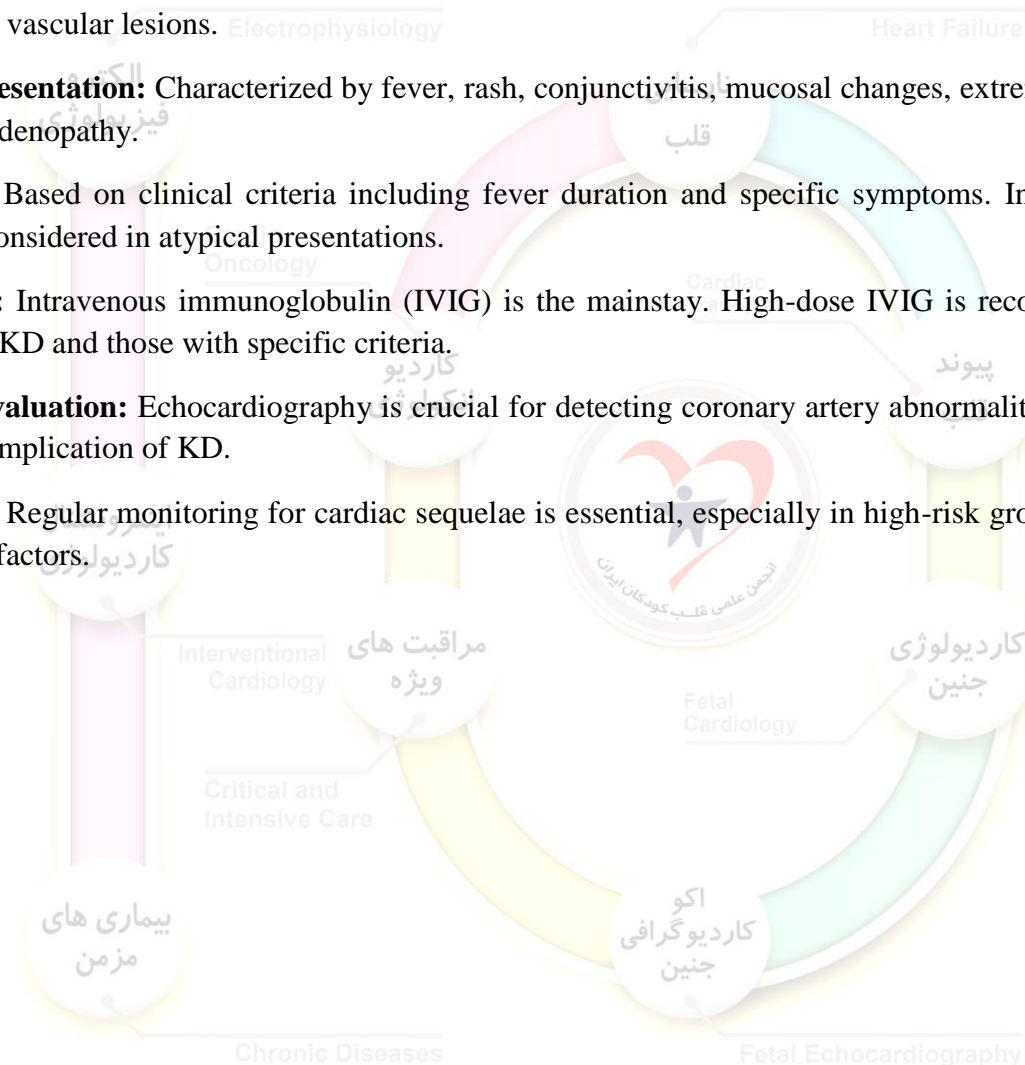
Clinical Presentation: Characterized by fever, rash, conjunctivitis, mucosal changes, extremity swelling, and lymphadenopathy.

Diagnosis: Based on clinical criteria including fever duration and specific symptoms. Incomplete KD should be considered in atypical presentations.

Treatment: Intravenous immunoglobulin (IVIG) is the mainstay. High-dose IVIG is recommended for incomplete KD and those with specific criteria.

Cardiac Evaluation: Echocardiography is crucial for detecting coronary artery abnormalities, which are the main complication of KD.

Follow-up: Regular monitoring for cardiac sequelae is essential, especially in high-risk groups identified by specific factors.



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Updates in Infective Endocarditis

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Infective endocarditis (IE) is an infection of the endocardium and/or heart valves, characterized by thrombus formation (vegetation) that may damage the endocardial tissue and valves. Although uncommon in children, IE carries significant morbidity and mortality.

1. Types of Infective Endocarditis:

- **Acute and Subacute Bacterial Endocarditis:** Caused by bacterial pathogens.
- **Nonbacterial Endocarditis:** Associated with viruses, fungi, and other microbiological agents.

2. Incidence Rates:

- Reported incidence rates of pediatric IE range from 0.3 to 3.3 per 100,000 individuals per year.
- The highest rates are observed in children with congenital heart disease (CHD).

3. Historical Trends:

- Rates of pediatric IE increased during the 20th century due to:
 - Dramatic improvements in survival among children with CHD.
 - Increased use of central venous catheters (CVC).

4. Risk Factors:

- **Congenital Heart Disease:**
 - Especially in children with cyanotic heart disease (35-60% of children with IE).
 - Rates increase with previous surgical interventions.
- **Central Venous Catheters (CVC):**
 - A significant risk factor.
- **Rheumatic Heart Disease:**
 - Remains relevant.
- **Other Risk Factors:**
 - Intravenous drug abuse (associated with right-sided IE).
 - Degenerative heart disease.
 - Recent hospitalization.
 - Hemodialysis.
 - Immunosuppression.

5. Microorganisms in IE:

- **Staphylococcus aureus (S. aureus):**
 - Most frequent causative agent, particularly in children without preexisting heart disease.
 - S. aureus-related IE is typically an acute, fulminant process with high mortality.
- **Gram-Negative Organisms:**
 - Patients with indwelling catheters are at risk for bacteremia.
 - However, gram-negative IE is rare due to poor adherence of these bacteria to the endocardium.
- **Neonatal IE:**
 - Likely etiologic agents include S. aureus, coagulase-negative staphylococci (CONS), Klebsiella pneumonia, and Enterobacter species.

This comprehensive review provides insights into the etiology, risk factors, and microbial profile of IE in pediatric patients.

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Cardiology Consultation for Non-cardiac Procedures

Fariba Alaei MD

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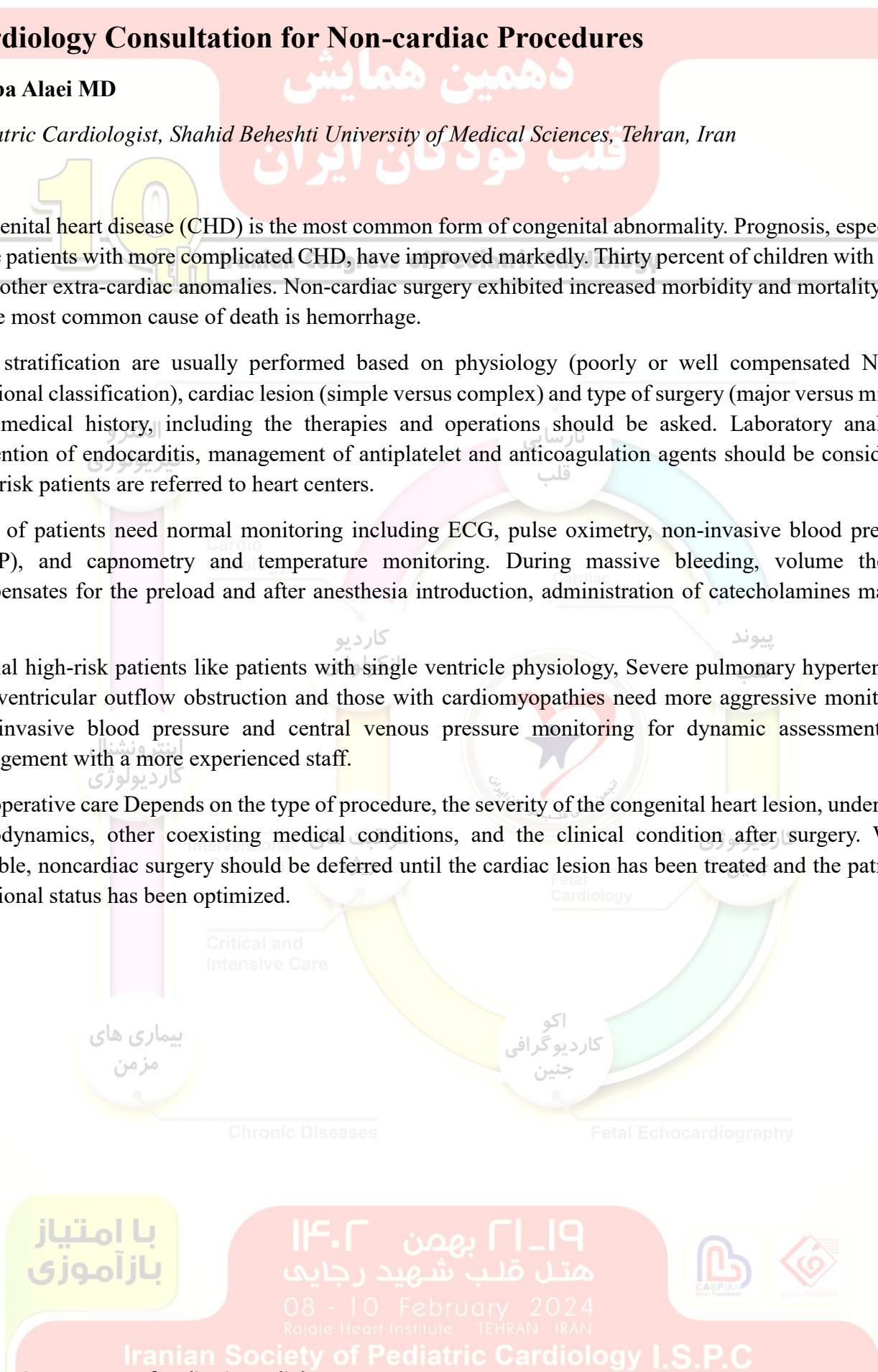
Congenital heart disease (CHD) is the most common form of congenital abnormality. Prognosis, especially in the patients with more complicated CHD, have improved markedly. Thirty percent of children with CHD have other extra-cardiac anomalies. Non-cardiac surgery exhibited increased morbidity and mortality. The single most common cause of death is hemorrhage.

Risk stratification are usually performed based on physiology (poorly or well compensated NYHA functional classification), cardiac lesion (simple versus complex) and type of surgery (major versus minor). Past medical history, including the therapies and operations should be asked. Laboratory analyses, prevention of endocarditis, management of antiplatelet and anticoagulation agents should be considered, high-risk patients are referred to heart centers.

Most of patients need normal monitoring including ECG, pulse oximetry, non-invasive blood pressure (NIBP), and capnometry and temperature monitoring. During massive bleeding, volume therapy compensates for the preload and after anesthesia introduction, administration of catecholamines may be used.

Special high-risk patients like patients with single ventricle physiology, Severe pulmonary hypertension, Left ventricular outflow obstruction and those with cardiomyopathies need more aggressive monitoring like invasive blood pressure and central venous pressure monitoring for dynamic assessment and management with a more experienced staff.

Postoperative care Depends on the type of procedure, the severity of the congenital heart lesion, underlying hemodynamics, other coexisting medical conditions, and the clinical condition after surgery. When possible, noncardiac surgery should be deferred until the cardiac lesion has been treated and the patient's functional status has been optimized.



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Infantile Hemangiomas and The Cardiac Relatives

Seyed Reza Miri MD

Pediatric Cardiologist

Infantile hemangiomas (IHs) represent the most common benign vascular tumors of infancy. Their natural evolution is well-established, characterized by an initial phase of proliferation followed by spontaneous involution over months to years. Despite this consistent course, there exists significant variation in clinical presentation, localization, life cycle, and associated conditions.

1. Clinical Variability:

- IHs can manifest at various extracutaneous sites, but they do not directly involve the heart.
- However, the heart may be indirectly affected:
 - Cardiac failure can develop in cases of multiple liver hemangiomas.
 - Diffuse neonatal hemangiomatosis may impact cardiac function.
 - Some systemic treatments for IHs may have cardiac side effects.

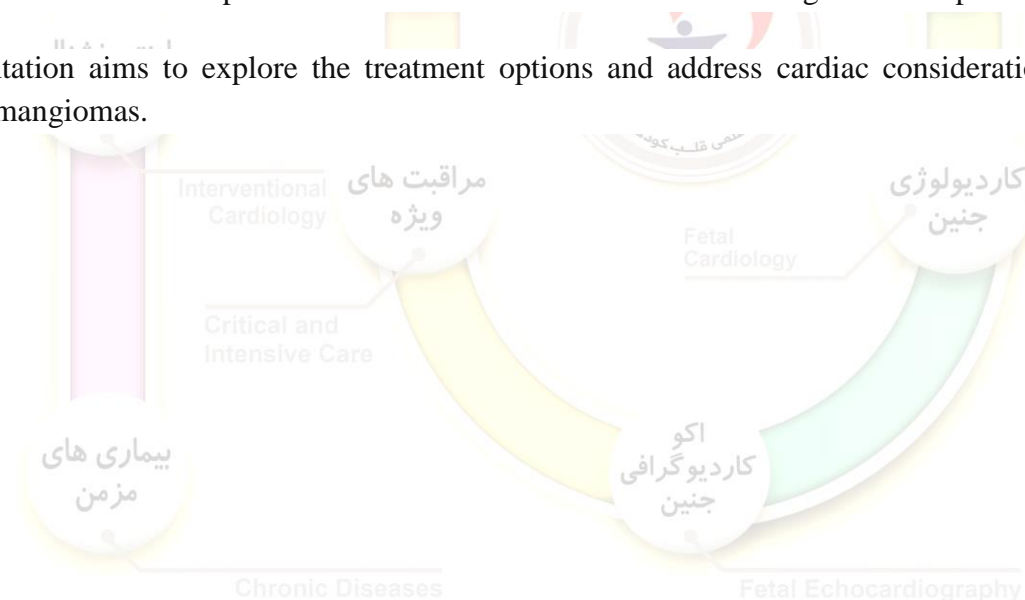
2. Cardiac Implications:

- Although IHs do not directly affect the heart, their systemic effects can lead to cardiac complications.
- Vigilance is necessary, especially in high-risk cases.

3. Treatment Considerations:

- Management strategies for IHs involve a multidisciplinary approach.
- Awareness of potential cardiac involvement is crucial during treatment planning.

This presentation aims to explore the treatment options and address cardiac considerations related to infantile hemangiomas.



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Fetal echocardiography in lung and thoracic anomalies

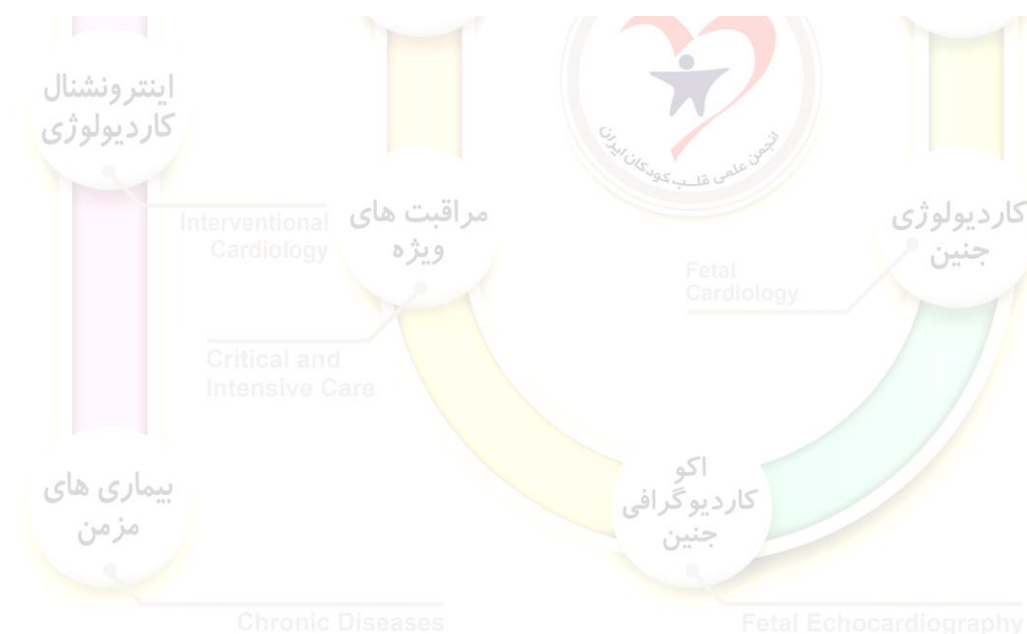
Alieh Nikdoost MD

Pediatric Cardiologist,

Background: In rare circumstances, fetuses can develop lung masses before birth. Two of the most common of these rare conditions are congenital pulmonary airway malformation (CPAM) and bronchopulmonary sequestration (BPS). Prenatal ultrasound detection of fetal anomalies has not only revolutionized management of the fetus and neonate, it has also changed our perception of the development and natural history of congenital anomalies throughout gestation. Changes in our understanding of fetal lung anomalies changed suggestion of termination of pregnancy (TOP) before fetal viability; in addition, some of these lesions may display dynamic changes in-utero. Congenital heart defect is one of the most defect and reports in 15-25% of patients. In addition, evaluation of cardiac function and hydrops in fetal echocardiography play an important role for decision making in this fetuses. Infants born with congenital diaphragmatic hernia (CDH) have an increased risk of congenital heart malformations about 12–25% of cases and survival rate for patients with major heart defect is low.

Result: We had two fetuses with large CPAM and cardiac displacement and normal cardiac anatomy and one fetus with CDH that aborted because of parental preference.

Conclusion: Nowadays treatments such as drainage procedure, fetal surgery and antenatal corticosteroids are used for improvement of prognosis. Meticulous evaluation maybe helpful in best decision making for patients.



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Pitfalls in pediatric echocardiography

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Echocardiography is still the most available and best diagnostic tool for cardiovascular diseases in children and adults. However several pitfalls may occur in the diagnosis of cardiovascular diseases in children with echocardiography. Sometimes the cause of these diagnostic mistakes is the limitations of echocardiography in diagnosis, but mostly the main reason is not using a regular and patient echocardiographic examination of the heart in all the views, along with an appropriate history and examination data of the patient. These pitfalls are not only related to very complex or rare diseases, and they may occur even in common and simple diseases such as ASD, VSD, COA and else.

ASD

Differentiating ASD from imaging caused by a simple dropout or PFO, although it seems very simple, is sometimes very difficult and challenging. Using different views, increasing the two-dimensional gain, using color and finally contrast echocardiography is useful in differentiating these cases. Diagnosing more complicated cases of ASD, especially the differentiation of ASD primum from the coronary sinus orifice, or the diagnosis of Sinus venosus ASDs and coronary sinus ASDs, are more difficult and requires more precision and experience.

VSD

Diagnosis of small or additional or misplaced VSDs, for example, muscular cases, especially apical ones, can be problematic. A complete evaluation and seeing of the apex of the heart in different views and the use of color and changing its gain can be helpful. Another problem is to recognize its concomitants, such as aortic valve prolapses or tricuspid pouch and its adhesions

COA

Diagnosis of coarctation is one of the most important responsibilities of pediatric cardiologists, which can easily be missed due to the presence of PDA or placement in an atypical and more distal location than usual or due to the child's non-cooperation in the proper position of the neck. PDA can cause disturbance in coarctation diagnosis due to the disturbance in aliasing detection of the coarctation site and the improvement and pseudo normalization of lower limb pulse and even abdominal aorta Doppler. Also, the presence of PDA can cause a false diagnosis of coarctation. A detailed examination of the arch of the aorta with accuracy and proper position of the neck, Doppler examination of the abdominal aorta and examination of the femoral pulse can be helpful.

Cardiac masses

Cardiac masses can be caused by tumors, clots, vegetations, or imaging caused by other cardiac structures such as Chiari's plexus, etc. It can be difficult to distinguish these cases from each other. Based on the history and previous history of the patient, the function of the heart and heart valve, and the location and nature of the mass, the type of mass can be differentiated to a great extent.

Aortic arch anomalies in the fetus

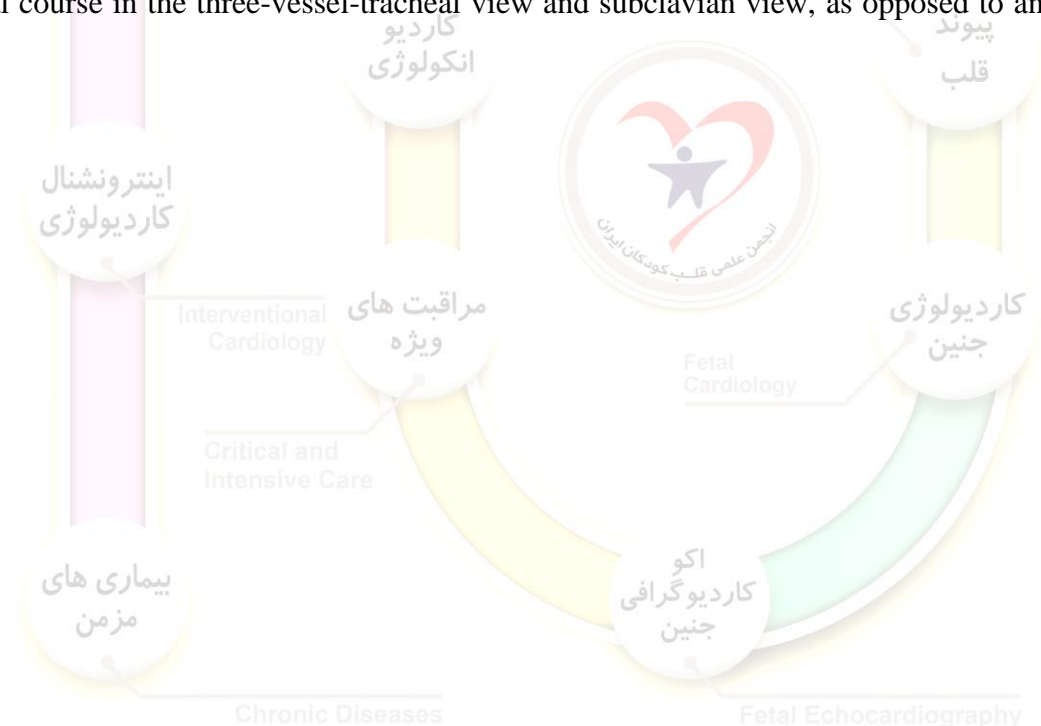
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Aortic arch anomalies result from abnormal branching or positioning of the aortic arch. There are four main categories of aortic arch anomalies identifiable during fetal life: right aortic arch, double aortic arch, circumflex aortic arch, and aberrant subclavian artery. Each anomaly presents with specific echocardiographic views. The three-vessel-tracheal view and subclavian views are crucial for diagnosing these anomalies. In cases of double aortic arch, circumflex retroesophageal aortic arch, and right aortic arch with left-sided ductus arteriosus connected to the proximal descending aorta, a "U" configuration appears instead of a "V" configuration in the three-vessel-tracheal view. A coronal scan of the aorta can differentiate between these anomalies. In cases of right aortic arch where the left-sided ductus arteriosus connects to the left innominate artery, the pulmonary artery and aortic arch exhibit a parallel alignment in the three-vessel-tracheal view. Conversely, in right aortic arch with a right-sided ductus arteriosus, a right-sided V-configuration is observed in the three-vessel-tracheal view, positioning both the aortic arch and the ductus arteriosus on the right side of the trachea. The identification of aberrant subclavian artery relies on its retrotracheal course in the three-vessel-tracheal view and subclavian view, as opposed to an antetracheal course.



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Patent Ductus Arteriosus Occluder Complications

Mehdi Ghaderian MD

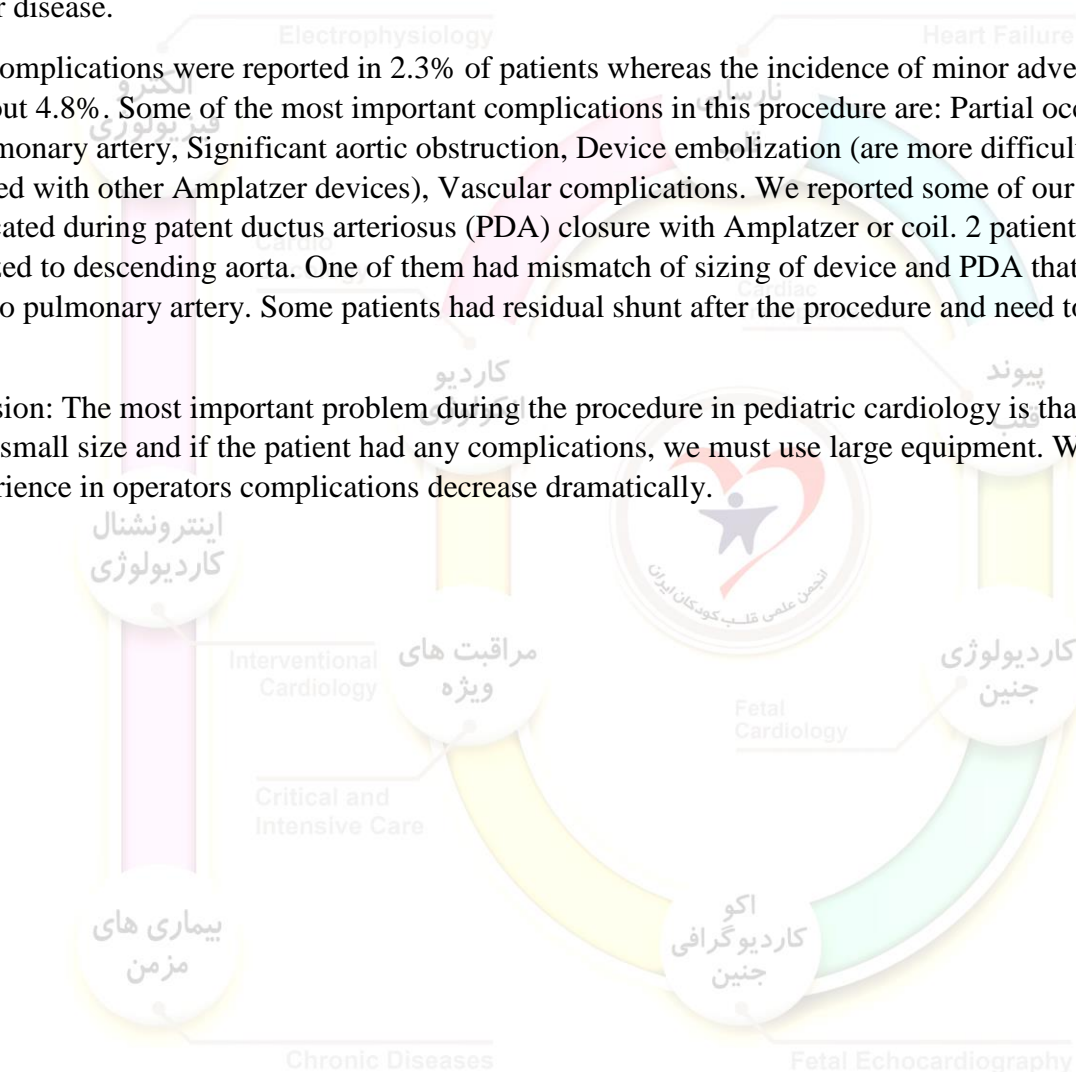
Pediatric Cardiovascular Research Center, Cardiovascular Research Institute, Isfahan University of Medical Sciences, Isfahan, Iran

Abstract:

Isolated patent ductus arteriosus (PDA) is one of the commoner congenital heart lesions comprising approximately 5-10% of congenital heart disease. It is recommended that all PDAs be closed to prevent infective endocarditis, relieve heart failure symptoms, and prevent progression to irreversible pulmonary vascular disease.

Major complications were reported in 2.3% of patients whereas the incidence of minor adverse events was about 4.8%. Some of the most important complications in this procedure are: Partial occlusion of the left pulmonary artery, Significant aortic obstruction, Device embolization (are more difficult to retrieve compared with other Amplatzer devices), Vascular complications. We reported some of our patients that complicated during patent ductus arteriosus (PDA) closure with Amplatzer or coil. 2 patients had device embolized to descending aorta. One of them had mismatch of sizing of device and PDA that pushed the device to pulmonary artery. Some patients had residual shunt after the procedure and need to use another device.

Conclusion: The most important problem during the procedure in pediatric cardiology is that the vessels are had small size and if the patient had any complications, we must use large equipment. With increasing of experience in operators complications decrease dramatically.



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Controversies in Large ASD Device Closure

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Introduction:

ASDs can be categorized into different types, including secundum ASDs, primum ASDs, sinus venosus ASDs, and coronary sinus type ASDs.

Patients with small or moderate ASDs are typically asymptomatic. However, individuals with large ASDs and significant shunts may experience exertional dyspnea or cardiac failure, often presenting in the fourth decade of life or later.

The management approach depends on the size and characteristics of the ASD. For ASDs smaller than 5 mm without volume overload (except in cases of paradoxical embolism), regular follow-up is recommended. Secundum ASDs larger than 5 mm but smaller than 38 mm, with a rim of 5-7 mm all around except towards the aorta, may be suitable for device closure. Larger ASDs, as well as associated tricuspid repair, sinus venosus defects, coronary sinus defects, or primum ASDs, typically require surgical intervention. The decision for closure is guided by factors such as pulmonary vascular resistance (PVR), systemic vascular resistance (SVR), and pulmonary artery pressure (PAP).

Closure of ASDs is indicated in two scenarios: 1) Symptomatic or hemodynamically significant shunt ($Q_p/Q_s > 1.5$ or evidence of right ventricular enlargement for body surface area), or 2) A small atrial defect with a history of paradoxical embolization resulting in stroke, transient ischemic attack, or peripheral embolism.

Challenges in Closure of Large ASDs:

Closing large ASDs can present several technical challenges, including deficient aortic rim, deficient posterior rim, deficient aortic and posterior rims, floppy rims, small child with a large ASD, unusually placed ASDs, multiple ASDs, and any ASD with a diameter exceeding 25mm. ASDs with absent inferior vena cava (IVC) or deficient IVC and posterior margins, as well as those larger than 36-38mm, are generally not suitable for device closure.

Techniques for Closure of Large ASDs:

Various techniques have been employed for the closure of large ASDs, including the Wahab technique, Hausdorf sheath, left upper pulmonary vein (LUPV) technique, balloon-assisted technique, right upper pulmonary vein (RUPV) technique, right Judkins catheter technique, and Boosfeld sheath.

Complication Avoidance:

During ASD closure procedures, it is important to be mindful of potential complications such as air embolism, secondary bleeding or hematoma formation, arrhythmias, device embolization, thromboembolism, erosions, and perforation.

In conclusion, the management of large ASDs remains a topic of debate and requires careful consideration of individual patient characteristics and technical challenges.

Unroofed Coronary Sinus with ASD Secundum and Interrupted IVC: A Case Report

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Abstract:

Iranian Congress of Pediatric Cardiology

We present a case report of an 8-year-old girl who was referred to our clinic due to cyanosis and mild shortness of breath during physical activity. The patient had a history of atrial septal defect (ASD) in infancy but did not receive proper follow-up care.

Clinical Findings:

Upon examination, the patient exhibited central cyanosis with an oxygen saturation level of 84%. Fingers clubbing was observed, and the patient had a grade II/VI ejectional systolic murmur.

Paraclinical Findings:

Electrocardiography (ECG) revealed a low right atrial (RA) rhythm and left axis deviation (LAD). Chest X-ray (CXR) showed mild cardiomegaly with slight enlargement of the pulmonary vascular markings (PVM).

Transthoracic echocardiography (TTE) demonstrated a small to moderately sized secundum ASD (measuring 9 mm) with left-to-right shunting. The coronary sinus (CS) appeared dilated, along with mild right atrial enlargement (RAE) and right ventricular enlargement (RVE). Additionally, mild dilation of the main pulmonary artery (MPA) and pulmonary annulus (PA) was noted, with a pulmonary artery flow gradient of 25 mm Hg. Tricuspid regurgitation (TR) was observed with a pressure gradient of 30 mm Hg, and pulmonary insufficiency (PI) was present with a pressure gradient of 15 mm Hg. The calculated Qp/Qs ratio was 1.6. A contrast-enhanced echocardiogram using agitated saline injected from the left arm confirmed the diagnosis of an unroofed coronary sinus (UCS).

Furthermore, computed tomography angiography (CTA) with contrast revealed an interrupted inferior vena cava (IVC) with hemiazygos continuation.

The patient was subsequently referred to a cardiac surgeon for total correction. Following the surgical intervention, the patient's oxygen saturation level increased to the normal range, and cyanosis resolved (O₂ sat: 97%). Postoperative follow-up is ongoing.

In conclusion, we present a rare case of unroofed coronary sinus with secundum ASD and interrupted IVC in an 8-year-old girl. Timely diagnosis and appropriate surgical intervention led to a successful outcome, with the resolution of cyanosis and improved oxygen saturation.

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Iranian Society of Pediatric Cardiology I.S.P.C

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First Iranian-Made Occluder

Hamid Amoozgar, Hamid Mohammadi, Jamal Arghavani hadi, Feryd Behroz

Shiraz university of Medical Sciences, Shiraz, Iran

Sharif University of technology, Tehran, Iran

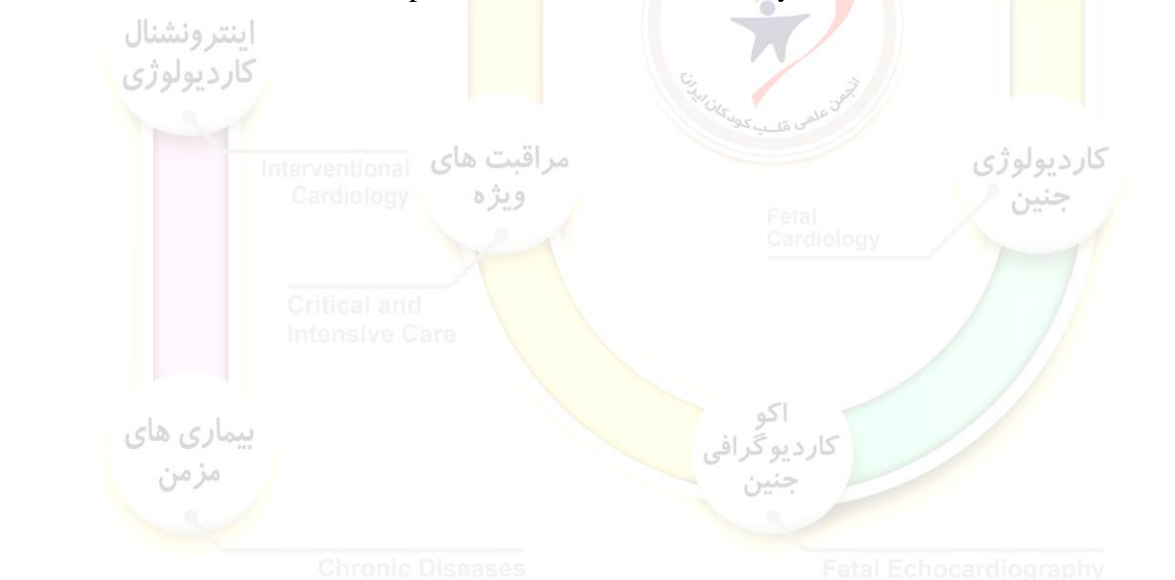
Abstract:

In recent years, advancements in technology in the field of cardiac and vascular repair have led to the development of various medical devices and tools. One such technology is the design and manufacture of occluders used for closing cardiac defects. In this study, the first Iranian-made occluder is evaluated.

This occluder has been fully designed and manufactured using advanced imaging and engineering technologies. Various evaluations, including laboratory studies and computer simulations, have been conducted on this occluder. The results of these evaluations have shown that this occluder performs very well in closing cardiac defects and has the capability of achieving satisfactory performance within the patients' bodies.

Furthermore, clinical studies on this occluder have been conducted in sheep, and very promising results have been presented in terms of reducing disease symptoms and improving quality of life.

Overall, the results of this research indicate that the Iranian-made occluder is an effective and high-quality option for closing cardiac defects and can significantly help improve the condition of patients. This study serves as a valuable starting point for the development and improvement of internal occluders in the country and can be fundamental to the development of the medical industry in Iran.



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Stenting of Pulmonary Artery Bifurcation

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Coronary angiography provides a formulated and coded assessment of stenosis at the bifurcation. In the case of the pulmonary artery, these stenoses can be divided into several categories: the first group involves stenosis in the distal main pulmonary artery (MPA). The technique used in these cases involves the use of a stent, two wires, and two balloons. During simultaneous inflation of the balloons on both sides, the stent is deployed distally with the support of the two wires. The second group includes stenosis in the proximal segments of both branches. In this case, the conventional technique of using two stents, two wires, and two balloons is employed, placed in a kissing configuration.

The third group involves stenosis both in the distal MPA and the proximal segments of both branches. In this situation, a creative technique is utilized, similar to the coronary angioplasty technique known as "Culotte," involving crush stenting on both sides. A desired stent and balloon can be used, and ultimately, the carina is completely stented with a double-layer stent before the bifurcation and a single-layer stent in the proximal branches. Another method in this category is the use of a specialized bifurcation stent, which is still under development and not yet widely available due to various production limitations similar to other stents.

The fourth group consists of stenosis in one branch, which can be completely resolved by placing a stent in the distal MPA and at the beginning of the affected branch. It is emphasized that the use of balloons with high-pressure tolerance and the ability to be longitudinally dissected in case of damage, such as the Atlas balloon, is essential for crushing.

It should be noted that in this presentation, the information from the CSI website was utilized.



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Cardiotoxicity in Leukemia: A Path to a Healthier Future

Dr. Golnar Mortaz Hejri,

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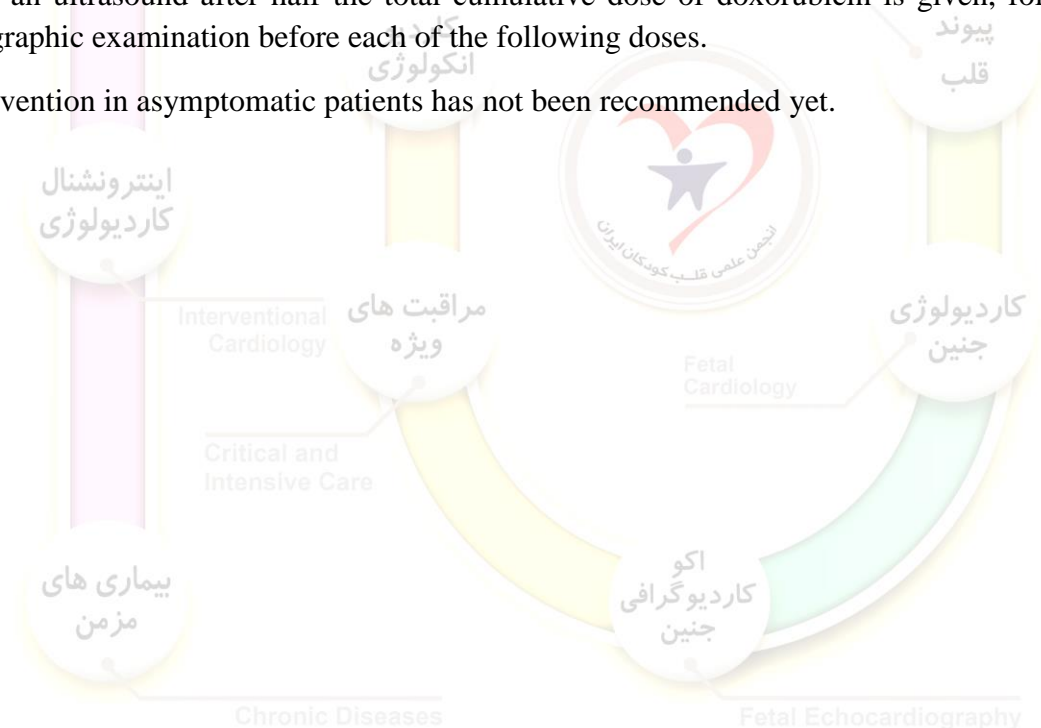
Chemotherapy is the main treatment method used in leukemia and consists of an association of several cytotoxic agents, showing an increased efficiency of up to 85% in inducing remission.

Cardiovascular side effects caused by chemotherapy are various, including arrhythmias, conduction disorders, heart failure (HF), acute coronary syndromes, myocarditis, and pericarditis. The most commonly encountered side effect is the alteration of left ventricular (LV) contractility, with the consequent decrease of its ejection fraction (LVEF).

Acute onset cardiotoxicity caused by an anthracycline is rare; it is defined as occurring within one week of administration of the anthracycline and is often reversible with discontinuation. Early onset chronic cardiotoxicity occurs within one year of administration, while late onset chronic cardiotoxicity presents greater than one year after administration of an anthracycline.

Monitoring patients with anthracycline includes echocardiography, ECG, and biomarkers. Most guidelines recommend an ultrasound after half the total cumulative dose of doxorubicin is given, followed by an echocardiographic examination before each of the following doses.

Primary prevention in asymptomatic patients has not been recommended yet.



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