CONGENITAL HEART DISEASES

Methodical elaborations for foreign students

МІНІСТЕРСТВО ОХОРОНИ ЗДОРОВ'Я УКРАЇНИ
харківський національний медичний університет

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INTRODUCTION

Congenital heart defect (CHD) may be defined as an anatomic malformation of the heart or great vessels which occurs during intrauterine development, irrespective of the age at presentation. Ventricular septal defect and coarctation of the aorta are typical examples of CHDs. In this chapter, a brief review of incidence, etiology and classification of CHD, and an overview of the most common congenital cardiac anomalies and their management will be presented.

Incidence of congenital heart defects.

Congenital heart defects are the most common type of birth defects, affecting approximately 1 in 125 live births. Although there have been many advances in the treatment of congenital heart defects, they remain a leading cause of infant mortality and can result in lifelong disability. CHD are serious and common conditions that have significant impact on morbidity, mortality, and healthcare costs in children and adults.

The reported incidence of congenital cardiac defects varies between 0.47 to 1.17 % of live births, but 0.6 % to 0.8 % of live births is considered typical. This would result in birth of 25,000 to 35,000 infants with CHD each year in the United States alone. Continental variations in birth prevalence have been reported, from 6.9 per 1000 births in Europe to 9.3 per 1000 in Asia. Congenital heart defects are more common than well-known congenital anomalies such as congenital pyloric stenosis, cleft lip, Down syndrome and congenital dislocation of the hip.

ETIOLOGY

CHD occur during the 2–8 weeks of fetal development. The exact cause of all congenital cardiac defects is not known. The majority of the defects can be explained by multifactorial inheritance hypothesis (Nora 1968) which states that a predisposed fetus, when exposed to a given environmental trigger (to which the fetus is sensitive) during the critical period of cardiac morphogenesis will develop the disease. This genetic and environmental interaction is most likely to be pathogenetic mechanism of congenital heart defects. Calculations based on this hypothesis predict the frequency of occurrence of the disease in first degree relatives to be square root of its frequency in the population; this fits the congenital heart disease figures (Nora 1968). A variety of factors have statistical association with certain heart defects and these may be termed risk factors. Maternal rubella appears to have causative association with heart defects. Significantly higher incidence of serologic evidence for Coxsackie B virus infection.

Among drugs, maternal ingestion of thalidomide during pregnancy is associated with high incidence heart defects in the offspring. Similar association

has been reported for some anticonvulsant drugs (particularly dilantin and trimethadione), alcohol (excessive), Lithium, sex hormones, diazepam, cartico-steroids, phenolhiazine, folic acid antagonists, cocaine and dextromethamphetamines. A higher incidence of cardiac abnormalities with maternal diabetes is well known. Gross chromosomal anomalies such as trisomy 21 (Down syndrome), trisomy D and E syndromes, Turner's syndrome (XO), partial deletion of chromosome 22 and cri-du-chat (partial dilation of the short arm of chromosome 5) are associated with a higher incidence of heart defects than normal population and are likely be responsible for the congenital heart defects. Some generalized syndromes, secondary to a single mutant gene (for example, Marfan) involving multiple organ systems are associated with cardiovascular defects peculiar to that particular syndrome (Rao 1977a). Both autosomal (dominant and recessive) and sex-linked (dominant and recessive) single mutant gene syndromes have been reported with CHD. Finally, less than 1% of congenital heart defects can be explained by simple Mendelian inheritance. Autosomal dominant transmission other than single mutant gene syndrome has been reported with atrial septal defect, patent ductus arteriosus, aortic stenosis, pulmonary stenosis, tetralogy of Fallot and hypertrophic cardiomyopathy. Autosomal recessive inheritance may be present in some forms of endocardial fibroelastosis. sex-linked transmission has not been reported with CHD.

However, recently questions have been raised as to the mitochondrial inheritance in which maternal transmission to almost all offspring occurs. In the presence of family history of congenital heart defect (parent or sibling) the probability of CHD in the offspring is higher than that seen in general population. In summary, the cause of congenital heart defects is largely unknown and the majority of them may be explained by multifactorial inheritance hypothesis. Extensive research on gene mapping that is currently in progress may unravel previously unknown genetic mechanisms for CHD.

Etiology. Main factors:

- Chromosomal disorders – 5 % (Syndromes of Down, Edwards, Turner, Patau);
- Mutation of one gene – 2–3 % (Syndromes of Marfan, Hurler, Kartagener, Holt Oram, Low etc);
- Factors of external environment – 1–2 % (infectious diseases, certain medications such as some antidepressants and opioid analgesics, smoking cigarettes during pregnancy, alcoholism of parents, ionizing and high frequency waves irradiation, chemical agents);
- Polygen-multifactorial inheritance – 90 % (heredity + factors of environment).

The risk of development CHD is higher if a sibling or a parent, especially the mother, has CHD – the absolute risk increasing from 1 % to 7–

10 %. While the causes of non-inherited CHD are still largely unknown, researchers have identified both genetic and noninherited *risk factors*:

- genetic or chromosomal abnormalities,
- environmental factors, such as maternal exposure to certain medications or organic solvents,
- maternal infections, such as rubella or influenza,
- maternal smoking during pregnancy,
- maternal diabetes,
- maternal overweight or obesity (women who are overweight or obese are 18-30% more likely to give birth to babies with CHD).

CLASSIFICATION OF CHD

Congenital heart disease occurs in 8 children for every 1000 live borns. Out of these 50% are significant in the sense that they produce haemodynamic effects. The classification of congenital heart diseases is shown in table 1. Congenital heart defects are classified into acyanotic and cyanotic depending upon whether the patients clinically exhibit cyanosis. The acyanotic defects may further be subdivided into obstructive lesions and left-to-right shunt lesions. The cyanotic defects, by definition, have right-to-left shunt.

Table 1. Classification of CHD

Acyanotic Congenital Heart Disease		Cyanotic Congenital Heart Disease	
with enriched pulmonary blood flow	obstruction to blood flow from ventricles	with diminished pulmonary blood flow	mixed blood flow
• Patent Ductus Arteriosus (PDA) • Atrial Septal Defect (ASD) • Ventricular Septal Defect (VSD) • Atrioventricular Septal Defect (AV Canal)	• Coarctation of the Aorta • Pulmonary Stenosis • Aortic Stenosis	• Fallot's disease, • Tricuspid atresia, • Ebstein's anomaly	• Transposition of great vessels with stenosis of the pulmonary artery, • Total anomalous pulmonary venous return, • Truncus arteriosus, • Hypoplastic left heart syndrome

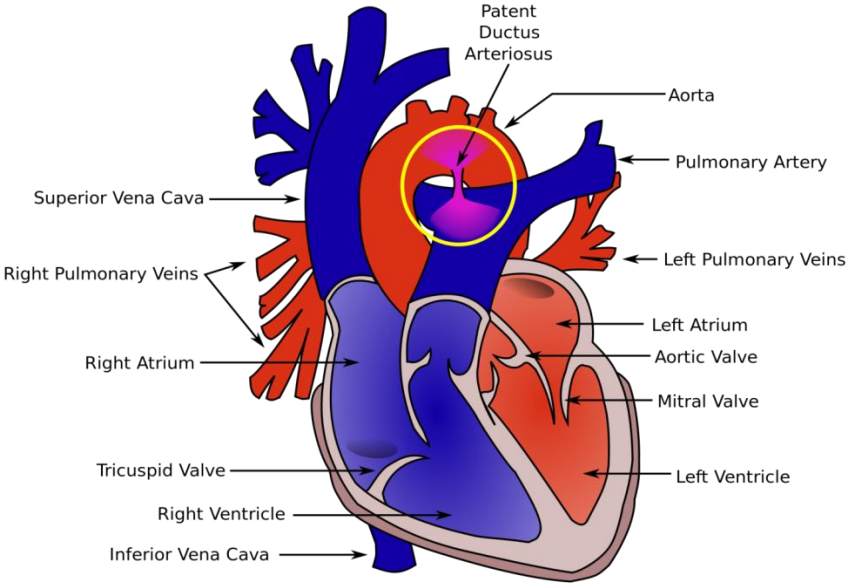
ACYANOTIC CONGENITAL HEART DISEASE

When there is a defect in the partition between left and right heart structures, the oxygenated blood is shunted from left-to-right because of generally lower pressure and/or resistance in the right heart than in the left. The physical findings are either a manifestation of flow across the defects or due to effects of excessive flow across the cardiac chambers (volume overload) and valves. The magnitude of the shunt determines the clinical presentation and symptoms.

Patent ductus arteriosus (PDA)

Patent ductus arteriosus (PDA) is one of the most common congenital heart defects. In healthy full-term infants, the ductus arteriosus closes within 48

to 72 hours. In premature infants born weighing more than 1,000 g, the ductus closes spontaneously in 67% by day 7 and in 94% by discharge. Overall, only 3 % of infants weighing more than 1,000 g may require intervention for a PDA.



The ductus arteriosus is a communication between the pulmonary artery and the aortic arch distal to the left subclavian artery. Patent ductus arteriosus (PDA) is the failure of the fetal ductus arteriosus to close after birth.

Ductus arteriosus, one of the fetal circulatory pathways, diverts the desaturated blood from the pulmonary artery into the descending aorta and placenta for oxygenation. After the infant is born, the ductus arteriosus constricts and closes spontaneously, presumably secondary to increased PO_2. But in some children, such spontaneous closure does not occur. This is more frequent in prematurely born infants. Patent ductus arteriosus (PDA) may be an isolated lesion and may be present in association with other defects. Isolated PDA constitutes 6 to 11 % of all CHDs.

Hemodynamics. As a result of higher aortic pressure, blood shunts Left-to-Right through the duct from Aorta to PA. Left to right shunting of blood through the patent ductus results in an increase in pulmonary blood flow. The amount of blood that flows through the ductus, and the degree of symptoms exhibited, is determined by the differences in systemic and pulmonary vascular resistance, and in the circumference and the length of the PDA.

NOTE: The PDA may be life-saving in infants with complex cyanotic heart defects or left sided obstructive defects, providing the only or major

source of pulmonary or systemic blood flow. A continuous PGE infusion will maintain the ductal opening and sustain life until cardiac surgery can be performed. PDA's may be present in premature infants.

Clinical Signs

Clinical presentation depends upon the size of the ductus. If the PDA is small, there are no symptoms and it is usually detected because of a murmur detected on a routine examination. Moderate to large ducti with large shunt may either present with symptoms of easy fatigability, symptoms associated congestive heart failure or respiratory symptoms suggestive of lung collapse (very large ductus in small babies).

The diagnosis is often suspected clinically, when an infant demonstrates signs of excessive shunting from the arterial to pulmonary circulation:

- continuous or systolic murmur; note, a "silent" PDA may also occur when the ductus shunt is large enough that nonturbulent flow fails to generate a detectible murmur;
- a low diastolic blood pressure (due to runoff into the ductus during diastole, more frequent in the most premature infants),
- a wide pulse pressure (due to ductus runoff or steal),
- hypotension (especially in the most premature infants),
- bounding pulses,
- increased serum creatinine concentration or oliguria,
- hepatomegaly.

Characteristic systolic-diastolic murmur at the base of the heart with maximum in the PA. This is classic continuous machine-like murmur. It localized to the 2nd left intercostal space or radiate down the left sternal border or to the left clavicle.

Signs of pulmonary edema are often seen, including tachypnea, decreased oxygen saturation, and increasing respiratory support. Chest radiography can show stigmata of pulmonary edema.

Chest x-ray

Chest film may show a normal-sized heart with normal pulmonary vascular markings with small ductus while cardiomegaly, increased pulmonary blood flow and left atrial enlargement may be seen with moderate to large ductus.

Electrocardiogram

The ECG may be normal or may show left atrial and left ventricular enlargement, depending upon the size of the ductus.

Echocardiogram

The echo may reveal varying degrees of left atrial and left ventricular enlargement, again depending upon the size of the ductus. The left ventricular contraction indices are normal unless severe myocardial dysfunction set in. *Doppler* echocardiography shows characteristic diastolic flow pattern in the

pulmonary artery, indicative of PDA. Characteristic color flow mapping distribution is also present

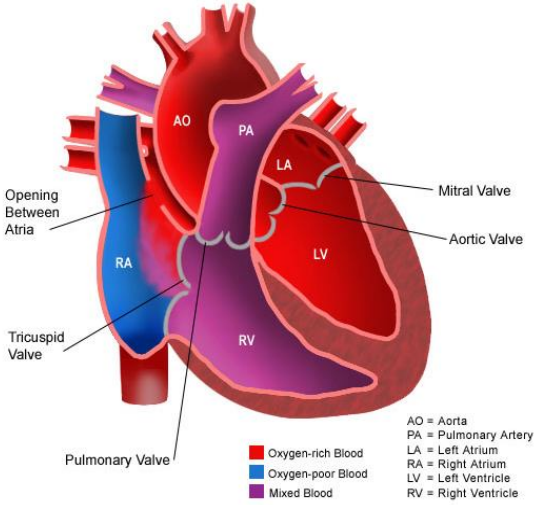
Management

It is generally believed that the presence of an isolated ductus is an indication for closure, mainly to prevent bacterial endocarditis. This can be performed at anytime, especially if associated with heart failure or pulmonary compromise. If the patient is asymptomatic, waiting until 6 to 12 months of age is generally recommended.

Until recently, surgical closure was the treatment of choice. While the risk of surgical closure is low, morbidity associated with it, namely anesthesia, endotrachial intubation and thoracotomy is universal. Because of this reason, less invasive, transcatheter closure techniques have been developed. These transcatheter methods are increasingly being used in closing PDAs.

Three pharmacologic treatments are available to induce constriction of a PDA: indomethacin, ibuprofen, and acetaminophen (paracetamol). Indomethacin and ibuprofen are classic nonsteroidal anti-inflammatory drugs (NSAIDs), which nonselectively inhibit the cyclooxygenase enzymes, preventing the conversion of arachidonic acid to prostaglandins, which play a central role in maintaining ductus patency. Since 1976, indomethacin has been used to treat PDA in premature infants.

Atrial septal defect (ASD) is a form of CHD that enables blood flow between the left and right atria via the interatrial septum (it is possible for blood to travel from the left side to the right side of the heart). Seen in 10–15 % cases of all CHD.



There are 4 anatomic types:

- Ostium primum - low in atrial septum, may involve a cleft mitral valve.
- Ostium secundum - center of the atrial septum. Most common type of ASD.
- Sinus venosus – high in the atrial septum. Associated with P-TAPVR.
- Coronary sinus - large opening between the coronary sinus and left atrium.

Physiology. Because of higher pressure in the left atrium, blood is usually shunted from the left atrium across the ASD and into the right atrium. ASD's are restrictive when they are small enough to provide resistance to flow across the septum. ASD's are non-restrictive when the opening is large enough that equal pressures occur in both atria. If the child has a cyanotic congenital heart defect, an ASD can provide an important shunt that allows mixing of oxygenated and venous blood within the atria. This may be necessary to sustain life.

Isolated ASD does not cause disorders of fetus blood circulation, and baby is born in time with normal body weight and normal sizes of heart. Isolated ASD's rarely cause symptoms during infancy. A small percentage of infants and children present with CHF and are treated with digoxin and diuretics. Symptoms are related to the size of the left-to-right shunt. Surgery is generally performed electively at 3–4 years of age. Very few ASD's close spontaneously after the 1st year of life.

Clinical Symptoms

Isolated ASD patients are usually asymptomatic and are usually detected at the time of preschool physical examination. Sometimes these defects are detected when echocardiographic studies are performed for some unrelated reason. A few patients do present with heart failure in infancy, although this is uncommon.

Physical examination

The right ventricular and right ventricular outflow tract impulses are increased and hyperdynamic. No thrills are usually felt. The second heart sound is widely split and fixed (splitting does not vary with respiration) and is the most characteristic sign of ASD. Ejection systolic clicks are rare with ASDs. The ejection systolic murmur of ASD is soft. The murmur is secondary to increased flow across the pulmonary valve and is heard best at the left upper sternal border. A mid-diastolic flow rumble is heard (with the bell of the stethoscope) best at the left lower sternal border. This is due to large volume flow across the tricuspid valve. There is no audible murmur because of flow across the ASD.

Diagnosis

Chest x-ray

Chest film usually reveals mild to moderate cardiomegaly, prominent main pulmonary artery segment and increased pulmonary vascular markings.

Electrocardiogram

The ECG shows mild right ventricular hypertrophy; the so-called diastolic volume overload pattern with rSR' pattern in the right chest leads.

Echocardiogram

Echocardiographic studies reveal enlarged right ventricle with paradoxical septal motion, particularly well-demonstrable on M-mode echocardiograms. By two-dimensional echocardiogram, the defect can be clearly visualized. In addition, demonstration of flow across the defect with color Doppler echocardiography is necessary to avid false positive studied.

Management

Despite lack of symptoms at presentation, closure of the ASD is recommended so as to 1) prevent development of pulmonary vascular obstructive disease later in life, 2) reduce chances for supra-ventricular arrhythmias and 3) prevent development of symptoms during adolescence and adulthood. Elective closure around age 4 to 5 years is recommended.

Closure during infancy is not undertaken unless the infant is symptomatic. Right ventricular volume overloading by echocardiogram and a Qp:Qs > 1.5 (if the child had cardiac catheterization) are indications for closure.

The conventional treatment of choice is surgical correction. While the secundum ASDs can be successfully repaired by open-heart surgical techniques with a low (< 1 %) mortality, the morbidity with cardiac surgery is universal, and residual scar is present in all. Because of this reason several transcatheter methods have been developed. But Ostium primum and sinus venosus defects are not amenable to transcatheter closure and surgical correction is the treatment of choice. In the ostium primum defect, apart from closing the ASD, the mitral valve should be repaired in such a manner as to preserve its function.

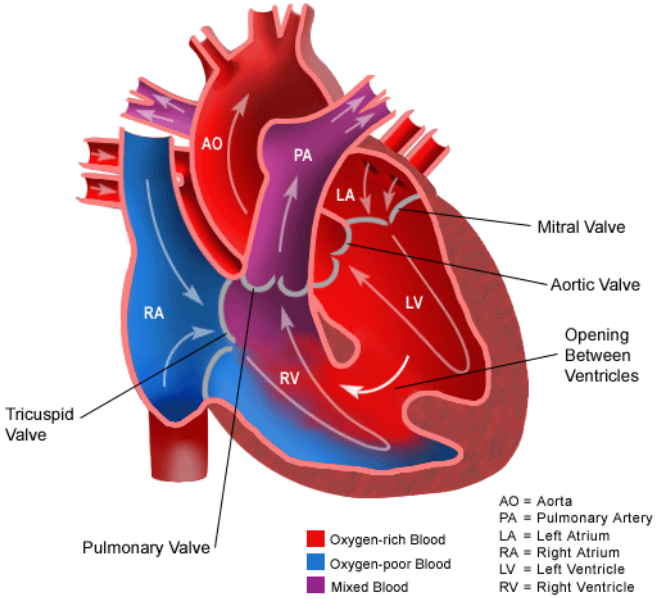
Ventricular septal defect (VSD)

Ventricular Septal Defect

A ventricular septal defect (VSD) is a communication (or multiple communications) between the right and left ventricles. VSD is the most common CHD and constitutes 20 % to 25 % of all CHDs. The defect may be small, medium or large and is classified based on its location in the inter-ventricular septum. There are 4 anatomic types:

- Perimembranous – upper portion of septum (most common).
- Subpulmonary – below pulmonary valve – and is called supracristal defect.

- Muscular – muscle portion of ventricular septum. Usually low in the ventricular septum. Multiple muscular defects may be referred to as ‘swiss cheese’ defects.
- Atrioventricular canal – located beneath the tricuspid valve. Also called an inlet VSD.



Physiology

Because of higher pressure in the left ventricle, blood is usually shunted from the left ventricle, across the VSD into the right ventricle, and into the pulmonary circulation. The risk of pulmonary hypertension depends on the size and location of the defect and the amount of pulmonary vascular resistance. Small VSD’s restrict the amount of blood shunting from left to right. Large VSD’s are non-restrictive, therefore a much higher degree of shunting occurs, and blood flow to the lungs is increased.

Identifying the type and size of the VSD is very important. The potential development of pulmonary hypertension needs to be followed closely to determine the timing of surgery. Children are often maintained on digoxin and diuretics for symptoms of CHF. These patients have an increased risk of developing pneumonia, and if a large VSD is present, they are at risk for development of subacute endocarditis. Surgery in early infancy may be recommended to prevent the development of pulmonary vascular disease.

Clinical Symptoms

The clinical symptomatology is largely dependent upon the size of the VSD. In small defects, the patients are usually asymptomatic and are detected because a cardiac murmur heard on routine examination. Patients with medium and large defects may present with symptoms of congestive heart failure (dyspnea, tachypnea, sweating and failure to gain weight) or with symptoms related to bronchial obstruction and/or respiratory infection.

Physical findings

These, again, depend upon the size of the defect. In small defects the only abnormality is a loud holosystolic murmur heard best at the left lower sternal border and is sometimes referred to as "maladie de Roger". In very small defects, murmur, though begins with first heart sound, may not last through the entire systole; the shorter the murmur, the smaller is the defect.

In medium and large defects, the right and left ventricular impulses are increased and somewhat hyperdynamic. A thrill may be felt at the left lower sternal border. The second heart sound is split unless there is pulmonary vascular obstructive disease, in which case a loud single second heart sound is heard. The pulmonary component of the second sound may be normal or increased, depending upon the degree of elevation pulmonary artery pressure.

The mid diastolic murmur may be heard at the apex. This murmur is due to increased flow across the mitral valve and usually indicates a Qp:Qs greater than 2 : 1.

Chest x-ray

The x-ray shows cardiomegaly and increased pulmonary vascular markings if the shunt is large. Left atrial enlargement may be noted.

Electrocardiogram

The ECG may be normal in very small defects or may show evidence for left ventricular hypertrophy in small to moderate defects while it may show biventricular or right ventricular hypertrophy in moderate to large defects. Electrocardiographic signs of left atrial enlargement may also be seen.

Echocardiogram Echo shows increase in left atrial and left ventricular size, which is again dependent upon the size of the VSD. The location and size of the VSD can be imaged by 2-dimensional echocardiography. Left-to-right shunting across the VSD can be demonstrated by Doppler echocardiography and color mapping.

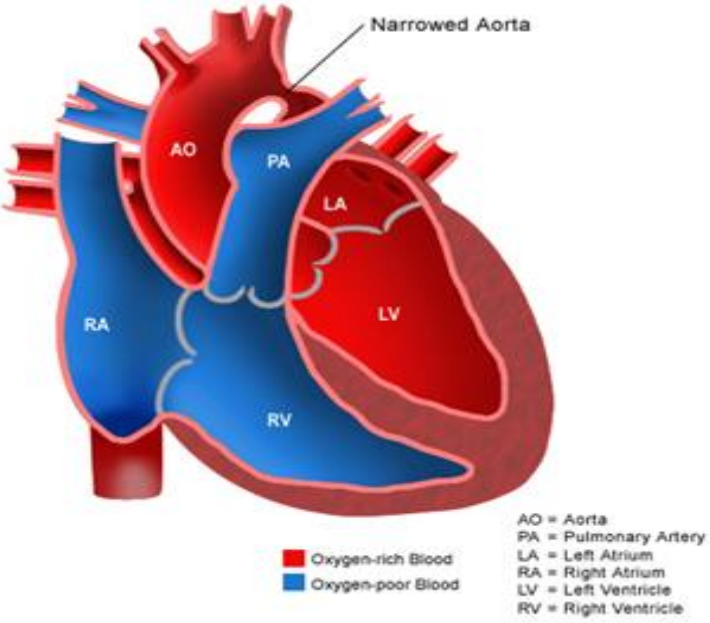
Management

The management strategies depend, to a large degree, on the size of the VSD. In small VSDs, reassurance of the parents, subacute bacterial endocarditis prophylaxis and periodic clinical follow-up are all that are necessary. In moderate-sized defects, treatment of heart failure, if present, should be undertaken. Failure to thrive and markedly enlarged left ventricle are probably

indications for surgical closure. In very large defects the heart failure should be treated aggressively. If the congestive heart failure is difficult to control with the usual anti-congestive measures or if failure to thrive is present, surgical closure should be undertaken. In large defects with near systemic pressures in the right ventricle and pulmonary artery, surgical closure should be performed prior to 18 to 24 months of age even if heart failure control and adequate weight gain are present. Total surgical correction is currently recommended. Transcatheter occlusion of VSD may be feasible in muscular defects and membranous defects with sufficient septum in the subaortic region so that the device can be implanted without interfering with aortic valve function

Large VSDs with severe elevation of pulmonary resistance (irreversible pulmonary vascular obstructive disease) are not candidates for surgery. Symptomatic treatment and erythropheresis for symptoms of polycthemia should be undertaken. These patients may eventually become candidates for lung transplantation.

Coarctation of the aorta



Coarctation of the aorta (CoA) is a narrowing in the aortic arch. Incidence of this defect is 7 % of all CHD. The coarctation may occur as a single lesion, as a result of improper development of the involved area of the aorta, or as a result of constriction of that portion of the aorta when the ductus arteriosus

constricts. The coarctation is most often located near the ductus arteriosus; if narrowing is proximal to the ductus it is 'pre-ductal', if it is distal to the ductus it is 'postductal'. Classic CoA is located in the thoracic aorta distal to the origin of the left subclavian artery, at about the level of the ductal structure. However, rarely, a coarcted segment may be present in the abdominal aorta.

Physiology

Aortic narrowing increases resistance to flow from the proximal to the distal aorta. As a result, pressure in the aorta proximal to the narrowing is increased and pressure in the aorta distal to the narrowing is decreased. Collateral circulation can develop in older children and adults to maintain adequate flow into the distal descending aorta.

Neonates and infants may present in shock when the ductus arteriosus closes. PGE must be started immediately to maintain ductal patency and surgery should be scheduled upon diagnosis. Some older children that present with CHF are maintained on digoxin and diuretics, and may be candidates for balloon dilation. Significant hypertension may also be seen, requiring anti-hypertensive therapy. A difference in blood pressure between upper and lower extremities helps identify this defect.

Clinical Symptoms

Children beyond infancy usually are asymptomatic; an occasional child will complain of pain or weakness in the legs. Most often, the coarctation is detected because of a murmur or hypertension which is detected on a routine examination.

Physical findings

A clinical diagnosis of CoA is best made by simultaneous palpation of femoral and brachial pulses. The left ventricular impulse may be increased. A thrill is usually felt in the supra-sternal notch. The first and second heart sounds are usually normal in isolated aortic coarctation. Since a large percentage (up to 60%) of patients with CoA have associated bicuspid aortic valves, an ejection systolic click may be heard at right upper and left mid sternal borders and apex; this click does not change with respiration. An ejection systolic murmur may be heard at left or right upper sternal borders, but is usually heard best over the back in the inter-scapular regions.

Palpation of the brachial and femoral artery pulses simultaneously will reveal delayed and decreased or absent femoral pulses. Blood pressure in both arms and one leg must be determined: a peak systolic pressure difference of more than 20 mmHg in favor of arms may be considered as evidence for coarctation of the aorta. Involvement of the left subclavian artery in the coarctation or anomalous origin of the right subclavian artery (below the level of coarctation) may produce decreased or absent left or right brachial pulses, respectively, and therefore palpation of both brachial pulses and measurement of blood pressure in both arms are important

Chest x-ray

Chest roentgenogram may show a normal sized heart or the heart may be mildly enlarged. Other roentgenographic features include a "3" sign on a highly penetrated chest x-ray, inverted "3" sign of the barium filled esophagus and rib-notching (secondary to collateral vessels).

Electrocardiogram

The ECG may be normal or may show left ventricular hypertrophy.

Echocardiogram

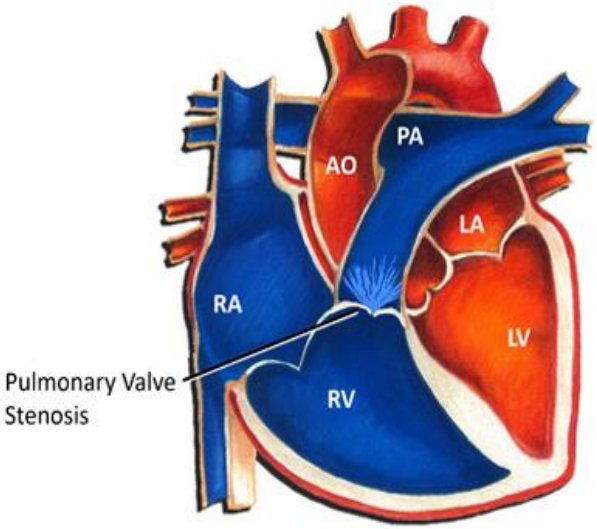
Echocardiographic studies usually reveal the coarctation in the supra-sternal notch, two-dimensional echo views of the aortic arch. Increased Doppler flow velocity in the descending aorta by continuous-wave Doppler and demonstrable jump in velocity at the coarcted segment by pulsed-Doppler technique are usually present. Extension of the Doppler flow signal into the diastole is indicative of significant obstruction.

Management

Significant hypertension and/or congestive heart failure are indications for intervention. In the presence of congestive heart failure, conventional anti-congestive measures including digitalis and diuretics should be promptly instituted. In the presence of hypertension, it is better to relieve the obstruction promptly rather than attempting to "treat" hypertension with antihypertensive drugs. Aortic coarctation may be relieved either by surgery or by balloon angioplasty. Symptomatic children should undergo relief of coarctation soon after the child is stabilized. Asymptomatic children should undergo the procedure electively. If neither hypertension nor heart failure is present, elective relief of the obstruction between the ages of 2 and 5 years is suggested. Waiting beyond 5 years is not advisable because of evidence for residual hypertension if the aortic obstruction is not relieved by the age of 5 years.

Pulmonary Stenosis

Pulmonary stenosis (PS) is a narrowing that obstructs blood flow from the right ventricle. Incidence of this defect is about 7–10 % of all CHD. It may be subvalvular, valvular, supravalvular or in the pulmonary arteries. Valvar stenosis is the most common type. When this presents in neonates, it is referred to as ‘critical pulmonary stenosis’.



Physiology

Pulmonary stenosis increases resistance to flow from the right ventricle. To maintain blood flow to the lungs, the right ventricle must generate higher pressures. The greater the pulmonary stenosis, the greater must be the pressure generated by the right ventricle. Because the pressure on the right side is higher, right ventricular hypertrophy is also present. Pulmonary stenosis may be mild, moderate or severe. When severe, the right ventricular hypertrophy may result in a right to left shunting through the foramen ovale.

Clinical Symptoms

Children with PS usually present with asymptomatic murmurs, although they can present with signs of systemic venous congestion (usually interpreted as congestive heart failure) due to severe right ventricular dysfunction or cyanosis because of right-to-left shunt across the atrial septum.

Physical findings

The right ventricular and the right ventricular outflow tract impulses are increased and a heave may be felt at the left lower and upper sternal borders. A thrill may be felt at the left upper sternal border and/or in the suprasternal notch. The first heart sound may be normal or loud. The second heart sound is variable, depending upon the degree of obstruction. An ejection systolic click is heard in most cases of valvar stenosis. The click is heard best at the left lower, mid and upper sternal borders and varies with respiration (decreases or disappears with inspiration). An ejection systolic murmur is heard best at the left upper sternal border and it radiates into infraclavicular regions, axillae and back.

Chest x-ray

In most cases, the chest film shows no cardiomegaly, but a characteristically dilated main pulmonary artery segment (post-stenotic dilatation) is visualized. The magnitude of pulmonary artery dilatation has no bearing on the severity of pulmonary valve stenosis.

Electrocardiogram

The ECG shows right ventricular hypertrophy; the degree of right ventricular hypertrophy is proportional to the severity of stenosis. Right atrial enlargement may be present.

Echocardiogram

The echo may show right ventricular enlargement without paradoxical septal motion and thickened and domed pulmonary valve leaflets. The Doppler flow velocity across the site of obstruction is increased and the magnitude of this increase reflects the severity of pulmonary valve stenosis.

Management

At the present time balloon pulmonary valvuloplasty is treatment of choice. The indications for intervention are similar to those prescribed for surgery: a peak-to-peak systolic pressure gradient > 50 mmHg across the pulmonary valve with a normal cardiac index.

CYANOTIC CONGENITAL HEART DISEASE

In cyanotic congenital heart defects systemic venous blood bypasses the pulmonary circulation and gets shunted across into the left side of the heart. Thus, there is systemic arterial desaturation. By definition, cyanotic congenital heart disease does not include cyanosis due to intrapulmonary right-to-left shunting and pulmonary venous desaturation secondary to congestive heart failure. There are usually multiple defects of the heart causing right-to-left shunt. Obstruction to pulmonary blood flow (for example tetralogy of Fallot), complete admixture of pulmonary and systemic venous returns (for example, total anomalous pulmonary venous return and double-inlet left ventricle) and parallel rather than in-series circulation (transposition of the great arteries) are the causes of right-to-left shunts and cyanosis.

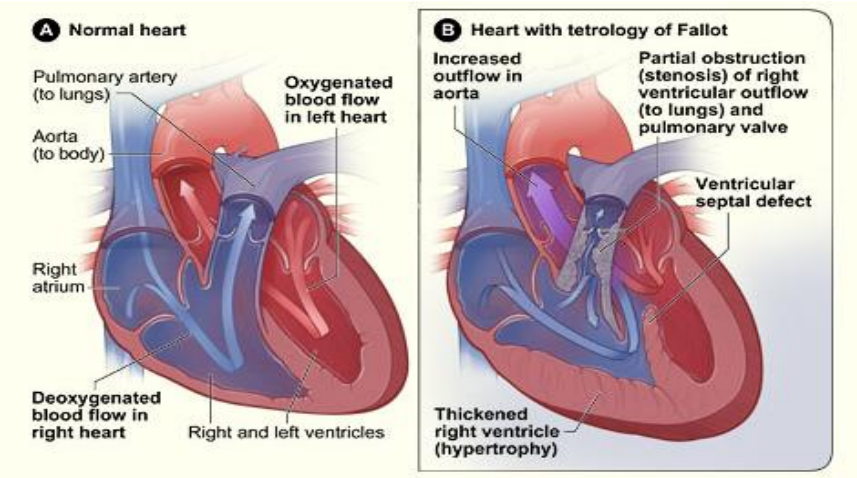
Tetralogy of Fallot

Tetralogy of Fallot (TOF) is the most common cause of cyanosis beyond one year of age and constitutes 10 % of all congenital heart defects. Fallot defined it as a constellation of four abnormalities to include a VSD, PS, right ventricular hypertrophy and dextroposition of the aorta (overriding aorta). There is a wide spectrum of right ventricular outflow tract obstruction (RVOTO) in TOF. It may be subvalvar, valvular and/or supravalvular. Typically, there is hypoplasia of the right ventricular outflow tract, stenosis of the pulmonary valve and hypoplasia of the pulmonary annulus and trunk. The right

and left pulmonary arteries are usually normal in size. Some infants with TOF may be referred to as a 'pink' TET, if no cyanosis is present.

Physiology

The hemodynamic changes and the degree of cyanosis that occur as a result of Tetrology of Fallot are directly proportional to the degree of subpulmonary stenosis (right ventricular outflow tract obstruction), and the resulting limitation to pulmonary blood flow. If RVOTO is mild, there is minimal shunting of blood from left to right across the VSD which may result in over circulation and signs of CHF. If severe RVOTO is present, a large amount of blood shunts from right to left, producing systemic arterial oxygen desaturation which can lead to severe hypoxemia and acidosis. As long as the ductus remains open, pulmonary blood flow is adequate.



Clinical Symptoms

The clinical presentation depends upon the degree of PS. With milder degrees of PS, symptoms may not be present until late childhood while with severe PS the presentation may be in the early infancy. Typically the infant may be pink (not cyanotic) as a neonate and develops cyanosis between 2 to 6 months of age. Most usual modes of presentation are asymptomatic murmur discovered on routine auscultation, bluish color (cyanosis) observed by the parent or primary physician, hypercyanotic spells, and decreased exercise tolerance. Hypercyanotic spells are variously described as anoxic spells, hypoxic spells, blue spells, paroxysmal dyspnea, paroxysmal hyperpnea and so no. The spells characteristically occur in tetralogy although they can be present in other lesions with similar physiology. They can occur any time between 1 month and 12 years of age but the peak incidence is 2 to 3 months. They can occur at any time of the day but most commonly seen after awakening from

sleep; crying, defecation and feeding are the common precipitating factors. Spells are characterized by increasing rate and depth of respiration (hyperpnea) with increasing cyanosis, progressing to limpness and syncope, occasionally terminating in convolutions, cerebrovascular accident or death. Spells may occur in tetralogy with mild arterial desaturation and conversely may not be present in patients with severe cyanosis.

NOTE: “TET Spells” or hypercyanotic spells are acute episodes of arterial oxygen desaturation secondary to intermittent worsening of right ventricular outflow tract obstruction causing right to left shunting across the VSD. The infant may become extremely irritable, cyanotic and may lose consciousness. Closely monitor for ‘TET spells’ which may lead to hypoxemia and acidosis. Symptoms depend on the degree of RVOTO and the amount of shunting.

Physical examination

Central cyanosis is observed in most cases of' tetralogy of Fallot. However, it should be noted that mild arterial desaturation may not cause clinically detectable cyanosis. Clubbing of fingers and toes is observed beyond the first few months of life. There are usually no signs of congestive heart failure. Prominent right ventricular impulse or heave may be present. A systolic thrill may be present at the left upper sternal border. The first heart sound may be normal or slightly increased. The second heart sound is single without an audible pulmonary component. A long ejection systolic murmur, caused by blood flow through the right ventricular outflow tract, is usually heard at the left upper sternal border. In contrast to PS with intact ventricular septum, the murmur of tetralogy becomes shorter and less intense with increasing severity of PS. During hypercyanotic spell the murmur disappears or becomes very soft.

Chest x-ray

On a chest roentgenogram the heart size is usually normal to minimally increased. An uplifted apex, thought to indicate right ventricular hypertrophy may be present and is described by some as "boot-shaped" heart. Concavity in the region of pulmonary conus, reflecting hypoplasia of the pulmonary outflow tract may be present. Pulmonary vascular markings are usually diminished. A right sided aortic arch may be present.

Electrocardiogram

The ECG shows signs of right ventricular hypertrophy. Right atrial enlargement is less commonly seen.

Echocardiogram

The echo is very helpful in confirming the diagnosis and in evaluating several of the issues related to TOF. Enlargement of the right ventricle, large VSD, aortic over-ride and right ventricular outflow tract obstruction can be imaged. Shunting across the VSD and increased Doppler flow velocity across the right ventricular outflow tract can be demonstrated.

Blood test

Hemoglobin and hematocrit along with red blood cell indices should be monitored periodically in all children with cyanotic congenital heart defects including TOF. The degree and duration of hypoxemia determine the level of hemoglobin. In the absence of adequate iron intake, relative anemia with hypochromia and microcytosis may develop. Because this is a risk factor for developing cerebrovascular accidents, the relative anemia should be treated with oral supplemental iron.

Management

The goal of management of TOF patients is to allow total surgical correction with minimal mortality and morbidity and to prevent or treat complications inherent to cyanotic heart defects in general and TOF in particular. Protection against subacute bacterial endocarditis, prevention and/or prompt treatment of dehydration, and periodic monitoring for relative anemia secondary to iron deficiency and prompt treatment when found should be undertaken.

Treatment of an infant with cyanotic spell may be summarized as follows:

1. The infant should be placed in a knee-chest position. The reason for its effectiveness appears to be related to its effect in increasing the systemic vascular resistance and thus decreasing the right-to-left shunt and improving the pulmonary flow.

2. Humidified oxygen via a facemask should be administered. Since the major defect in the spell syndrome is pulmonary oligemia rather than alveolar hypoxia, oxygen administration has limited usefulness. If the infant is unduly disturbed by the facemask, oxygen therapy may be discontinued.

3. Morphine sulfate, 0.1 mg/kg subcutaneously, may be effective in aborting the spell. The mechanism of action is not clearly delineated, but its depressive effect on the central nervous system respiratory drive (thus reducing hyperpnea) and sedation of the infant may be important.

4. Once the physical examination is completed (and the limited but important laboratory studies are obtained) the infant should be left undisturbed and allowed to rest; this in itself may improve the infant's condition.

5. Correction of metabolic acidosis (with sodium bicarbonate), anemia (by blood transfusion), and dehydration (by appropriate fluids), if present, is very important at this stage.

6. If the spell continues, vasopressors to increase the systemic vascular resistance and thus increase the pulmonary blood flow may be tried. Methoxamine 20-40 mg in 250 ml of 5% dextrose in water may be administered intravenously;

7. Alternatively, propranolol, 0.1 mg/kg body weight, diluted in 50 ml of 5% dextrose in water, may be slowly administered intravenously while

monitoring the heart rate (by ECG if possible). Should there be marked bradycardia, propranolol should be stopped. Once it is found to be effective, the infant may be switched to oral propranolol 1-4 mg/kg/day in three and four divided doses. The mechanism of action of propranolol include negative inotropic effect on the right ventricular infundibular myocardium, prevention of decrease in systemic vascular resistance and/or prevention of ventilatory response (hyperpnea) to hypoxia, all through beta adrenergic blockade.

8. Infrequently, general anesthesia may be necessary to break the spell.

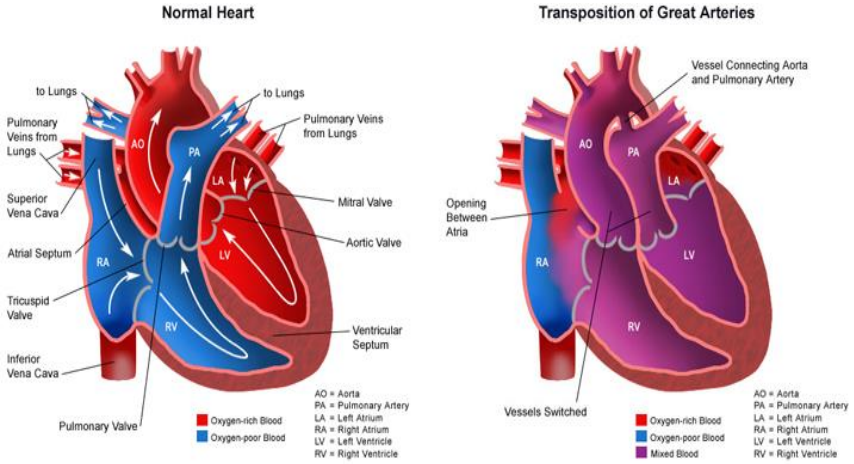
9. If the infant does not improve with any of the aforementioned measures, an emergency systemic-to-pulmonary artery shunt should be performed. Occasionally, total correction, if the anatomy is adequate, may he performed on an emergency basis. The important principle is that the infant requires more pulmonary blood flow.

If the infant improves with the management outlined above, total surgical correction of the cardiac defects, if anatomically feasible, or a systemic-to-pulmonary artery shunt to improve pulmonary blood flow on an elective basis within the next day or so may be performed.

Cyanotic Congenital Heart Disease with mixed blood flow

Transposition of the Great Arteries

Transposition of the great arteries (TGA) is the most common cyanotic congenital heart defect presenting in the newborn period. It constitutes 5 % of all CHD and 10 % of all cyanotic CHD.



In Transposition of the Great Arteries (TGA) the aorta arises from the anatomic right ventricle and the pulmonary artery arises from the anatomic left ventricle. The most common form of transposition occurs when the ventricles are normally positioned and the aorta is malposed anteriorly and rightward above the right ventricle. A VSD is present in 40 % of patients with TGA. Abnormal coronary artery patterns present in 33 % of cases.

Physiology

Blood flows from the RA to the RV and out through the aorta, carrying deoxygenated blood to the body. Blood flows from the LA to the LV and out through the PA, carrying oxygenated blood to the lungs. This results in two separate, parallel circulations that require mixing at the atrial, ventricular or ductus arteriosus level. The degree of desaturation present will depend primarily on the amount of mixing between systemic and pulmonary venous blood. The extent of inter-circulatory mixing in TGA depends on the number, size and position of the anatomic communications.

Clinical Symptoms

Clinical features depend upon the anatomic type, namely:

Group I, TGA with intact ventricular septum;

Group II, TGA with VSD, and

Group III, TGA with VSD and PS.

In group I with intact septum, the infants usually present with cyanosis within the first week of life. They may otherwise be asymptomatic. However, they will soon become tachypnoeic and develop respiratory distress. If they are not appropriately treated, they become acidotic and go on to become lethargic without lack of spontaneous movement, and eventually die.

Group II TGA patients with VSD present with symptoms of congestive heart failure (tachypnea, tachycardia, sweating, and poor feeding) between 4 to 8 weeks of life, but the cyanosis is minimal.

Group III patients (TGA with VSD and PS) have variable presentation, depending upon the severity of PS. If there is poor mixing, they may present early in life and mimic TGA with intact septum. If the PS is severe, the presentation is essentially similar to that described in the TOF section. With moderate PS the presentation is late with longer survival. With mild PS, congestive heart failure signs may be present, similar to group II patients.

Physical examination

The group I patients with intact septum are usually severely cyanotic but are without distress until severe hypoxemia and acidosis develop. Clubbing is not present in the newborn period and may not develop until 3 to 6 months. The right ventricular impulse is increased and the second heart sound is single. Either no murmur or a nonspecific ejection systolic murmur may be auscultated.

In group II patients, tachypnea, tachycardia, minimal cyanosis, hepatomegaly, increased right and left ventricular impulses, single second

sound, a holosystolic murmur at the left lower sternal border and a mid-diastolic flow rumble (murmur) at the apex may be present.

In group III patients, the findings are similar to TGA with intact septum, TGA with VSD, or TOF depending upon the degree of mixing and severity of PS.

Chest x-ray in the intact septum group is benign with normal to minimal cardiomegaly and normal to slightly increased pulmonary vascular marking. The thymic shadow may rapidly involute and a narrow pedicle (superior mediastinum) may be seen. A combination of the above signs may sometimes assume "egg-shaped" appearance on a postero-anterior chest roentgenogram. In group II patients with VSD, moderate to severe cardiomegaly and increased pulmonary vascular markings are usually seen. In group III patient, mild to at worst moderate cardiomegaly may be observed. The pulmonary vascular marking may be increased, normal or decreased, dependent upon the severity of PS.

Electrocardiogram

The ECG in a neonate with TGA and intact septum (Group I) may be normal with the usual right ventricular preponderance seen during this age. In older infants clear-cut right ventricular hypertrophy is seen and in addition right atrial enlargement may be observed. In group II patients, biventricular hypertrophy and left atrial enlargement are usual. In group III, right ventricular or biventricular enlargement is seen.

Echocardiogram

The echo is usually helpful in the diagnosis and assessment. Demonstration of transposition of the great arteries is somewhat difficult in view of the fact that atrial and ventricular anatomy is normal and the aortic and pulmonary valves look similar on echocardiographic study. If one can follow the great vessel arising from the left ventricle and demonstrate its bifurcation, identifying it as a pulmonary artery, the diagnosis is easy. The presence of an inter-atrial communication and patent ductus arteriosus and shunt across them by color and pulsed Doppler can also be evaluated. In addition to these, demonstration of VSD and PS will place the patients into the respective groups.

Other laboratory data Blood gas values are useful in demonstrating the degree of hypoxemia and ventilatory status. Hemoglobin and hematocrit are particularly useful in the follow-up of older children.

Management

Untreated, TGA with intact septum carries a poor prognosis. The initial management of this and other cyanotic neonates is similar. Monitoring the infant's temperature and maintenance of neutral thermal environment is extremely important. In hypoxemic infants, ambient oxygen should be administered. In cyanotic CHD patients, no more than 0.4 FIO2 is necessary. Metabolic acidosis (pH < 7.25), if any, should be corrected with sodium bicarbonate (usually 1–2 mEq/kg diluted half and half with 5 % or 10 %

dextrose solution) immediately. Respiratory acidosis should be cared for by appropriate suctioning, intubation and assisted ventilation. Hypoglycemia may be a significant problem; therefore, the infant's serum glucose should be monitored and the neonates should routinely receive 10% dextrose in water intravenously. If hypoglycemia (< 30 mg/100 ml) occurs, 15 % to 20 % dextrose solution should be administered. Similarly hypocalcemia should be monitored for and treated, if found. If an infant is getting progressively hypoxemic, it is likely that the intercirculatory pathways (patent foramen ovale and patent ductus arteriosus) are closing. Prostaglandin El (PGEI) (0.05 to 0.1 mcg/kg/min) intravenously may help open the ductus, thus improve oxygenation. Balloon atrial septostomy may be necessary to improve hypoxemia even after PGE1. Total surgical correction by arterial switch procedure is the treatment of choice in these neonates.

PREVENTING BIRTH DEFECTS

The American Heart Association guidelines to help prospective mothers lower the risk of CHD in their babies urges them to:

- Take a multivitamin with folic acid daily.
- Get rubella and flu shots and avoid contact with people with fever-related illnesses.
- Obtain preconception and prenatal care, with specific attention to detection and effective management of diabetes.
- Discuss prescription and over-the-counter medication use with a doctor.
- Avoid exposure to organic solvents.
- Stop smoking
- Avoid drinking alcohol while pregnant
- Stop use of any illegal or "street" drugs
- Check with doctor to make sure any medication (over-the-counter or prescription) is safe to take during pregnancy.

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Навчальне видання

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