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CONGENITAL HEART DISEASES IN CHILDREN

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ВРОЖДЕННЫЕ ПОРОКИ СЕРДЦА У ДЕТЕЙ

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Учебно-методическое пособие

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PREVA[illegible]CE

Congenital heart diseases (CHD) are the most common group of structural malformations in children which occurs in 0.5–0.8 % of live births. The incidence is higher in stillborns (3–4 %), abortuses (10–25 %), and premature infants (about 2 % excluding patent ductus arteriosus [PDA]). This overall

incidence does not include mitral valve prolapse, PDA of preterm infants, and bicuspid aortic valves (present in 1–2 % of adults).

CHD have a wide spectrum of severity in infants: about 2–3 in 1,000 newborn infants will be symptomatic in the 1st yr of life. The diagnosis is established by 1 wk of age in 40–50 % of patients with CHD and by 1 mo of age in 50–60 % of patients. With advances in both palliative and corrective surgery in the last 20 yrs, the number of children with CHD surviving to adulthood has increased dramatically. Despite these advances, CHD remains the leading cause of death in children with congenital malformations.

RELATIVE FREQUENCY OF THE MOST COMMON CONGENITAL HEART DISEASES (CHD)

1. Ventricular septal defect 25–30 %.
2. Atrial septal defect (secundum) 6–8 %.
3. Patent ductus arteriosus 6–8 %.
4. Coarctation of aorta 5–7 %.
5. Tetralogy of Fallot 5–7 %.
6. Transposition of great arteries 3–5 %.
7. Others 5–10 %.

Most CHD are well tolerated in the fetus because of the parallel nature of the fetal circulation. Even the most severe CHD (hypoplastic left heart syndrome) can usually be well compensated by the fetal circulation. In this example, the entire fetal cardiac output would be ejected by the right ventricle via the ductus arteriosus into both the descending and ascending aortae (the latter filling in a retrograde fashion). It is only after birth when the fetal pathways (ductus arteriosus and foramen ovale) are closed that the full hemodynamic impact of an anatomic abnormality becomes apparent. One notable exception is the case of severe regurgitant lesions, most commonly of the tricuspid valve. In these lesions (Ebstein anomaly), the parallel fetal circulation cannot compensate for the volume load imposed on the right side of the heart. In utero heart failure, often with fetal pleural and pericardial effusions, and generalized ascites (nonimmune hydrops fetalis) may occur.

Although the most significant transitions in circulation occur in the immediate perinatal period, the circulation continues to undergo changes after birth, and these later changes may also have a hemodynamic impact on cardiac lesions and their apparent incidence. As pulmonary vascular resistance falls over the 1st several weeks of life, left-to-right shunting through intracardiac defects increases and symptoms become more apparent. Thus, in patients with a ventricular septal defect (VSD), heart failure is often manifested between 1 and 3 mo of age.

The severity of various defects can also change dramatically with growth; some VSDs may become smaller and even close as the child ages. Alternatively, stenosis of the aortic or pulmonary valve, which may be mild in the newborn period, may become worse if valve orifice growth does not keep pace with patient growth. The physician should always be alert for associated congenital malformations, which can adversely affect the patient's prognosis.

ETIOLOGY OF CHD

The cause of most CHD is unknown, but rapid progress is being made in identifying the genetic basis of many congenital heart lesions. **Most cases of CHD** were thought to be **multifactorial** and result from **a combination of genetic predisposition and environmental stimulus**. A small percentage of CHD were related to chromosomal abnormalities, in particular, trisomy 21, 13, and 18 and Turner syndrome: heart disease is found in more than 90 % of patients with trisomy 18, 50 % of patients with trisomy 21, and 40 % of those with Turner syndrome. Other genetic factors were suspected to play a role in CHD; for example, certain types of VSDs (supracristal) are more common in Asian children. The risk of recurrence of CHD increases if a 1st-degree relative (parent or sibling) is affected.

A growing list of CHD have been associated with **specific chromosomal abnormalities**, and several have even been linked to specific gene defects.

A well-characterized genetic cause of CHD is the deletion of a large region of chromosome 22q11, known as the DiGeorge critical region. The estimated prevalence of 22q11 deletions is 1 in 4,000 live births. Cardiac

lesions associated with 22q11 deletions are most often seen in association with either the DiGeorge syndrome or the Shprintzen (velocardiofacial) syndrome.

The acronym CATCH 22 has been used to summarize the major components of these syndromes (*c*ardiac defects, *a*bnormal facies, *t*hymic aplasia, *c*left palate, and *h*ypocalcemia). The specific cardiac anomalies are conotruncal defects (tetralogy of Fallot, truncus arteriosus, double-outlet right ventricle, subarterial VSD) and branchial arch defects (coarctation of the aorta, interrupted aortic arch, right aortic arch). Congenital airway anomalies such as tracheomalacia and bronchomalacia are sometimes present. Although the risk of recurrence is extremely low in the absence of a parental 22q11 deletion, it is 50 % if one of the parents carries the deletion.

Two to 4 % of cases of CHD are associated with known **environmental or adverse maternal conditions and teratogenic influences**, including maternal diabetes mellitus, phenylketonuria, or systemic lupus erythematosus; congenital rubella syndrome or other viruses; and maternal ingestion of drugs (lithium, ethanol, warfarin, thalidomide, antimetabolites, anticonvulsant agents).

Parents who have a child with CHD require genetic counseling regarding the probability of a cardiac malformation occurring in subsequent children. With the exception of syndromes known to be due to mutation of a single gene, most CHD is still relegated to a multifactorial inheritance pattern, which should result in a low risk of recurrence. The incidence of CHD in the normal population is approximately 0.8 %, and this incidence increases to 2–6 % for a 2nd pregnancy after the birth of a child with CHD or if a parent is affected. This recurrence risk is highly dependent on the type of lesion in the 1st child. When two 1st-degree relatives have CHD the risk for a subsequent child may reach 20–30 %.

Fetal echocardiography (EchoCG) improves the rate of detection of CHD in high-risk patients. The resolution and accuracy of fetal EchoCG are not perfect, and families should be counseled that a normal fetal EchoCG does not guarantee the absence of CHD. CHD may also evolve during the course of the pregnancy; for example, moderate aortic stenosis with a normal-sized left ventricle at 18 wk of gestation may evolve into aortic atresia with a hypoplastic left ventricle by

34 wk because of decreased flow through the atria, ventricle, and aorta during the latter half of gestation.

When structural abnormalities of other systems are present, consider echocardiography for associated cardiac disorders.

EVALUATION OF THE INFANT OR CHILD WITH CHD

The initial evaluation for suspected CHD involves a systematic approach **with 3 major components**.

1. CHD can be divided into 2 major groups based on the presence or absence of cyanosis, which can be determined by physical examination aided by pulse oximetry.

2. These 2 groups can be further subdivided according to whether the chest X-ray shows evidence of increased, normal, or decreased pulmonary vascular markings.

3. The electrocardiogram (ECG) can be used to determine whether right, left, or biventricular hypertrophy exists. The character of the heart sounds and the presence and character of any murmurs further narrow the differential diagnosis. The final diagnosis is then confirmed by EchoCG or cardiac catheterization, or by both.

The most common CHD:

1. ***Acyanotic*** — ventricular septal defect (VSD), PDA, atrial septal defect (ASD).

2. ***Outflow obstruction*** — pulmonary stenosis, aortic stenosis, coarctation of the aorta.

3. ***Cyanotic*** — tetralogy of Fallot, transposition of the great vessels, AV septal defect complete

ACYANOTIC CONGENITAL HEART DISEASE: THE LEFT-TO-RIGHT SHUNT LESIONS

PATENT DUCTUS ARTERIOSUS

During fetal life, most of the pulmonary arterial blood is shunted through the ductus arteriosus into the aorta. Functional closure of the ductus normally occurs soon after birth, but if the ductus remains patent when pulmonary vascular resistance falls, aortic blood is shunted into the pulmonary artery. The aortic end of the ductus is just distal to the origin of the left subclavian artery, and the ductus enters the pulmonary artery at its bifurcation. Female patients with PDA outnumber males 2:1. PDA is also associated with maternal rubella infection during early pregnancy. It is a common problem in premature infants, where it can cause severe hemodynamic derangements and several major sequelae.

When a term infant is found to have a PDA, the wall of the ductus is deficient in both the mucoid endothelial layer and the muscular media. In a premature infant, the PDA usually has a normal structure; patency is the result of hypoxia and immaturity. Thus, a PDA persisting beyond the 1st few weeks of life in a term infant rarely closes spontaneously or with pharmacologic intervention, whereas if early pharmacologic or surgical intervention is not required in a premature infant, spontaneous closure occurs in most instances. A PDA is seen in 10 % of patients with other CHD and often plays a critical role in providing pulmonary blood flow when the right ventricular outflow tract is stenotic or atretic or in providing systemic blood flow in the presence of aortic coarctation or interruption.

Pathophysiology. As a result of the higher aortic pressure, blood shunts left to right through the ductus, from the aorta to the pulmonary artery. The extent of the shunt depends on the size of the ductus and on the ratio of pulmonary to systemic vascular resistance. In extreme cases, 70 % of the left ventricular output may be shunted through the ductus to the pulmonary circulation. If the PDA is small, pressure within the pulmonary artery, the right ventricle, and the right atrium is normal. However, if the PDA is large, pulmonary artery pressure may be elevated to systemic levels during both systole and diastole. Patients with a large PDA are at extremely high risk for the development of pulmonary vascular disease if left unoperated. Pulse

pressure is wide because of runoff of blood into the pulmonary artery during diastole.

Clinical Manifestations. A small PDA does not usually have any symptoms associated with it. A large PDA will result in heart failure similar to that encountered in infants with a large VSD. ***Retardation of physical growth may be a major manifestation in infants with large shunts.***

A large PDA will result in striking physical signs attributable to the wide pulse pressure, most prominently, bounding peripheral arterial pulses. The heart is normal in size when the ductus is small but moderately or grossly enlarged in cases with a large communication. The apical impulse is prominent and, with cardiac enlargement, is heaving. A thrill, maximal in the 2nd left interspace, is often present and may radiate toward the left clavicle, down the left sternal border, or toward the apex. It is usually systolic but may also be palpated throughout
the cardiac cycle. The classic continuous murmur is described as being like machinery or rolling thunder in quality. It begins soon after onset of the 1st sound, reaches maximal intensity at the end of systole, and wanes in late diastole. It may be localized to the 2nd left intercostal space or radiate down the left sternal border or to the left clavicle. When pulmonary vascular resistance is increased,
the diastolic component of the murmur may be less prominent or absent. In patients with a large left-to-right shunt, a low-pitched mitral mid-diastolic murmur may be audible at the apex as a result of the increased volume of blood flow across the mitral valve.

Diagnosis. If the left-to-right shunt is small, the *ECG* is normal; if the ductus is large, left ventricular or biventricular hypertrophy is present. The diagnosis of an isolated, uncomplicated PDA is untenable when right ventricular hypertrophy is noted.

Radiographic studies in patients with a large PDA show a prominent pulmonary artery with increased intrapulmonary vascular markings. Cardiac size depends on the degree of left-to-right shunting; it may be normal or moderately

to markedly enlarged. The chambers involved are the left atrium and ventricle. The aortic knob is normal or prominent.

The *EchoCG* of the cardiac chambers is normal if the ductus is small. With large shunts, left atrial and left ventricular dimensions are increased. The size of the left atrium is usually quantitated by comparison to the size of the aortic root, known as the LA:Ao ratio. Scanning from the suprasternal notch allows direct visualization of the ductus. Color and pulsed Doppler examinations demonstrate systolic or diastolic (or both) retrograde turbulent flow in the pulmonary artery and aortic retrograde flow in diastole.

The clinical pattern is sufficiently distinctive to allow an accurate diagnosis by noninvasive methods in most patients. In patients with atypical findings or when associated cardiac lesions are suspected, *cardiac catheterization* may be indicated. It demonstrates normal or increased pressure in the right ventricle and pulmonary artery, depending on the size of the ductus. The presence of oxygenated blood shunting into the pulmonary artery confirms a left-to-right shunt. The catheter may pass from the pulmonary artery through the ductus into the descending aorta. Injection of contrast medium into the ascending aorta shows opacification of the pulmonary artery from the aorta and identifies the ductus.

Prognosis and Complications. Patients with a small PDA may live a normal span with few or no cardiac symptoms, but late manifestations may occur. Spontaneous closure of the ductus after infancy is extremely rare. ***Cardiac failure*** most often occurs in early infancy in the presence of a large ductus but may occur late in life even with a moderate-sized communication. The chronic left ventricular volume load is less well tolerated with aging.

Infective endarteritis may be seen at any age. Pulmonary or systemic emboli may occur. Rare complications include aneurysmal dilatation of the pulmonary artery or the ductus, calcification of the ductus, noninfective thrombosis
of the ductus with embolization, and paradoxical emboli. Pulmonary hypertension (Eisenmenger syndrome) usually develops in patients with a large PDA who do not undergo surgical treatment.

Treatment. Irrespective of age, patients with PDA require surgical or catheter closure. In patients with a small PDA, the rationale for closure is prevention of bacterial endarteritis or other late complications. In patients with a moderate to large PDA, closure is accomplished to treat heart failure or prevent the development of pulmonary vascular disease, or both. Once the

diagnosis
of a moderate to large PDA is made, treatment should not be unduly postponed after adequate medical therapy for cardiac failure has been instituted.

Surgical closure of PDA can be accomplished by thoracoscopic techniques to minimize scarring and reduce postoperative discomfort. Because the case fatality rate with surgical treatment is considerably less than 1 % and the risk without it is greater, ligation and division of the ductus are indicated in asymptomatic patients, preferably before 1 yr of age. Pulmonary hypertension is not a contraindication
to surgery at any age if it can be demonstrated at cardiac catheterization that the shunt flow is still predominantly left to right and that severe pulmonary vascular disease is not present. After closure, symptoms of frank or incipient cardiac failure rapidly disappear.

Infants who had failed to thrive usually have immediate improvement in physical development. The pulse and blood pressure return to normal, and the machinery-like murmur disappears. The radiographic signs of cardiac enlargement and pulmonary overcirculation disappear over a period of several months, and the ECG becomes normal.

Transcatheter PDA closure is routinely performed in the cardiac catheterization laboratory. Small PDAs are generally closed with intravascular coils. Moderate to large PDAs may be closed with a catheter-introduced sac into which several coils are released or with an umbrella-like device.

ATRIAL SEPTAL DEFECTS (ASDS)

ASDs can occur in any portion of the atrial septum (secundum, primum, or sinus venosus), depending on which embryonic septal structure has failed to develop normally. Less commonly, the atrial septum may be nearly absent, with the creation of a functional single atrium. Isolated secundum ASDs account for ~7 % of CHD. The majority of cases of ASD are sporadic; however, autosomal dominant inheritance does occur as part of the Holt-Oram syndrome (hypoplastic or absent radii, 1st-degree heart block, ASD).

An isolated valve-incompetent *patent foramen ovale (PFO)* is a common EchoCG finding during infancy. It is usually of no hemodynamic significance and is not considered an ASD; however, a PFO may play an important role if other structural heart defects are present. If another cardiac anomaly is causing

increased right atrial pressure (e. g., pulmonary stenosis or atresia, tricuspid valve abnormalities, right ventricular dysfunction), venous blood may shunt across
the PFO into the left atrium with resultant cyanosis. Because of the anatomic structure of the PFO, left-to-right shunting is unusual outside the immediate newborn period. In the presence of a large volume load or a hypertensive left atrium (e. g., secondary to mitral stenosis), the foramen ovale may be sufficiently dilated to result in a significant atrial left-to-right shunt. A valve-competent but probe-patent foramen ovale may be present in 15–30 % of adults. An isolated PFO does not require surgical treatment, although it may be a risk for paradoxical (right to left) systemic embolization.

Atrioventricular Septal Defects (Ostium Primum and Atrioventricular Canal or Endocardial Cushion Defects)

The abnormalities encompassed by AV septal defects are grouped together because they represent a spectrum of a basic embryologic abnormality, a deficiency of the AV septum. An *ostium primum defect* is situated in the lower portion of the atrial septum and overlies the mitral and tricuspid valves. In most instances, a cleft in the anterior leaflet of the mitral valve is also noted. The tricuspid valve is usually functionally normal, although some anatomic abnormality of the septal leaflet is generally present. The ventricular septum is intact.

An *AV septal defect*, also known as an AV canal defect, consists of contiguous atrial and ventricular septal defects with markedly abnormal AV valves. The severity of the valve abnormalities varies considerably; in the complete form of AV septal defect, a single AV valve is common to both ventricles and consists of an anterior and a posterior bridging leaflet related to the ventricular septum, with a lateral leaflet in each ventricle. The lesion is common in children with Down syndrome and may occasionally occur with pulmonary stenosis.

Pathophysiology. The basic abnormality in patients with *ostium primum defects* is the combination of a left-to-right shunt across the atrial defect and mitral insufficiency. The shunt is usually moderate to large, the degree of mitral insufficiency is generally mild to moderate, and pulmonary arterial pressure is typically normal or only mildly increased. The physiology of this lesion is therefore similar to that of an ostium secundum ASD.

In *AV septal defects*, the left-to-right shunt occurs at both the atrial and ventricular levels. Additional shunting may occur directly from the left ventricle to the right atrium because of absence of the AV septum. Pulmonary hypertension and an early tendency to increase pulmonary vascular resistance are common. AV valvular insufficiency increases the volume load on one or both ventricles. Some right-to-left shunting may also occur at both the atrial and ventricular levels and lead to mild but significant arterial desaturation. With time, progressive pulmonary vascular disease increases the right-to-left shunt so that clinical cyanosis develops (Eisenmenger physiology).

***Clinical Manifestations*.** Many children with ostium primum *defects* are asymptomatic, and the anomaly is discovered during a general physical examination. In patients with moderate shunts and mild mitral insufficiency, the physical signs are similar to those of the secundum ASD, but with an additional apical murmur caused by mitral insufficiency.

A history of exercise intolerance, easy fatigability, and recurrent pneumonia may be obtained, especially in infants with large left-to-right shunts and severe mitral insufficiency. Cardiac enlargement is moderate or marked, and
the precordium is hyperdynamic. Auscultatory signs produced by the left-to-right shunt include a normal or accentuated 1st sound; wide, fixed splitting of the 2nd sound; a pulmonary systolic ejection murmur sometimes preceded by a click; and a low-pitched, mid-diastolic rumbling murmur at the lower left sternal edge or apex, or both, as a result of increased flow through the AV valves. Mitral insufficiency may be manifested by an apical harsh (occasionally very high pitched) holosystolic murmur that radiates to the left axilla.

With *complete AV septal defects*, congestive heart failure and intercurrent pulmonary infection usually appear in infancy. During these episodes, minimal cyanosis may be evident. The liver is enlarged and the infant shows signs of failure to thrive. Cardiac enlargement is moderate to marked, and a systolic thrill is frequently palpable at the lower left sternal border. A precordial bulge and lift may be present as well. The 1st heart sound is normal or accentuated. The 2nd heart sound is widely split if the pulmonary flow is massive. A low-pitched, mid-diastolic rumbling murmur is audible at the lower left sternal border, and
a pulmonary systolic ejection murmur is produced by the large pulmonary flow.

The harsh apical holosystolic murmur of mitral insufficiency may also be present.

***Diagnosis*.** Chest *X-ray* in complete AV septal defects shows marked cardiac enlargement caused by the prominence of both ventricles and atria. The pulmonary artery is large, and pulmonary vascularity is increased.

The *ECG* in patients with a complete AV septal defect is distinctive. The principal abnormalities are (1) superior orientation of the mean frontal QRS axis with left axis deviation to the left upper or right upper quadrant, (2) counterclockwise inscription of the superiorly oriented QRS vector loop, (3) signs of biventricular hypertrophy or isolated right ventricular hypertrophy, (4) right ventricular conduction delay (RSR' pattern in leads V_3R and V_1), (5) normal or tall P waves, and (6) occasional prolongation of the P-R interval.

The *EchoCG* shows signs of right ventricular enlargement with encroachment of the mitral valve echo on the left ventricular outflow tract; the abnormally low position of the AV valves results in a "gooseneck" deformity of the left ventricular outflow tract on both echocardiography and angiography.

In normal hearts, the tricuspid valve inserts slightly more toward the apex than the mitral valve does. In AV septal defects, both valves insert at the same level because of absence of the AV septum. In complete AV septal defects, the ventricular septal echo is also deficient and the common AV valve is readily appreciated. *Color flow Doppler EchoCG* demonstrates left-to-right shunting at the atrial, ventricular, or ventricular-to-atrial levels and semiquantitate the degree of AV valve insufficiency.

Cardiac catheterization and *angiocardiography* may be required to confirm the diagnosis, although most patients can be operated on without catheterization.

It demonstrates the magnitude of the left-to-right shunt, the severity of pulmonary hypertension, the degree of elevation of pulmonary vascular resistance, and the severity of insufficiency of the common AV valve. Children with ostium primum defects generally have normal or only moderately elevated pulmonary arterial pressure. Conversely, complete AV septal defects are associated with right ventricular and pulmonary hypertension and, in older patients, with increased pulmonary vascular resistance.

***Prognosis and Complications*.** The prognosis for complete AV septal defects depends on the magnitude of the left-to-right shunt, the degree of elevation of pulmonary vascular resistance, and the severity of AV valve insufficiency. Death from cardiac failure during infancy used to be frequent before the advent of early corrective surgery. In patients who survived without surgery, pulmonary vascular obstructive disease or, more rarely, pulmonic stenosis usually developed. Most patients with ostium primum defects and minimal AV valve involvement are asymptomatic or have only minor, nonprogressive symptoms until they reach the 3rd–4th decade of life, similar to the course of patients with secundum ASDs.

Treatment. Ostium primum defects are approached surgically from an incision in the right atrium. The cleft in the mitral valve is located through the atrial defect and is repaired by direct suture. The defect in the atrial septum is usually closed by insertion of a patch prosthesis. The surgical mortality rate for ostium primum defects is low. Surgical treatment of complete AV septal defects is more difficult, especially in infants with cardiac failure and pulmonary hypertension. Because of the risk of pulmonary vascular disease developing as early as 6–12 mo of age, surgical intervention must be performed during infancy. Correction of these defects can be accomplished in infancy, and palliation with pulmonary arterial banding is reserved for the subset of patients who are either too small or have other associated lesions that make early corrective surgery too risky. The atrial and ventricular defects are patched and the AV valves reconstructed.

Complications include surgically induced heart block requiring placement of a permanent pacemaker, excessive narrowing of the left ventricular outflow tract requiring surgical revision, and eventual worsening of mitral regurgitation requiring replacement with a prosthetic valve.

VENTRICULAR SEPTAL DEFECT

VSD is the most common cardiac malformation and accounts for 25 % of CHD. Defects may occur in any portion of the ventricular septum, but most are of the membranous type. These defects are in a posteroinferior position, anterior to the septal leaflet of the tricuspid valve. VSDs between the crista supraventricularis and the papillary muscle of the conus may be associated with pulmonary stenosis and other manifestations of the tetralogy of Fallot. VSDs

superior to the crista supraventricularis (supracristal) are less common; they are found just beneath the pulmonary valve and may impinge on an aortic sinus and cause aortic insufficiency. VSDs in the midportion or apical region of the ventricular septum are muscular in type and may be single or multiple (Swiss cheese septum).

Pathophysiology. The physical size of the VSD is a major, but not the only determinant of the size of the left-to-right shunt. The level of pulmonary vascular resistance in relation to systemic vascular resistance also determines the shunt's magnitude. When a small communication is present (usually < 0.5 cm^2), the VSD is called *restrictive* and right ventricular pressure is normal. The higher pressure in the left ventricle drives the shunt left to right; the size of the defect limits the magnitude of the shunt. In large *nonrestrictive* VSDs (usually > 1.0 cm^2), right and left ventricular pressure is equalized. In these defects, the direction of shunting and shunt magnitude are determined by the ratio of pulmonary to systemic vascular resistance.

After birth in patients with a large VSD, pulmonary vascular resistance may remain higher than normal, and thus the size of the left-to-right shunt may initially be limited. As pulmonary vascular resistance continues to fall in the 1st few weeks after birth because of normal involution of the media of small pulmonary arterioles, the size of the left-to-right shunt increases. Eventually, a large left-to-right shunt develops, and clinical symptoms become apparent. In most cases during early infancy, pulmonary vascular resistance is only slightly elevated, and the major contribution to pulmonary hypertension is the extremely in large pulmonary blood flow. However, in some infants with a large VSD, pulmonary arteriolar medial thickness never decreases. With continued exposure of the pulmonary vascular bed to high systolic pressure and high flow, pulmonary vascular obstructive disease develops. When the ratio of pulmonary to systemic resistance approaches 1:1, the shunt becomes bidirectional, the signs of heart failure abate, and the patient becomes cyanotic (Eisenmenger physiology).

Clinical Manifestations. The clinical findings of patients with a VSD vary according to the size of the defect and pulmonary blood flow and pressure. *Small VSDs* with trivial left-to-right shunts and normal pulmonary arterial

pressure are the most common. These patients are asymptomatic, and the cardiac lesion is usually found during routine physical examination. A loud, harsh, or blowing holosystolic murmur is present and heard best over the lower left sternal border, and it is frequently accompanied by a thrill. A short, harsh systolic murmur localized to the apex in a neonate is often a sign of a tiny muscular VSD.
In the immediate neonatal period, the left-to-right shunt may be minimal because of higher right-sided pressure, and therefore the systolic murmur may not be audible during the 1st few days of life. In premature infants, the murmur may be heard early because pulmonary vascular resistance decreases more rapidly.

Large VSDs with excessive pulmonary blood flow and pulmonary hypertension are responsible for dyspnea, feeding difficulties, poor growth, profuse perspiration, recurrent pulmonary infections, and cardiac failure in early infancy. Cyanosis is usually absent, but duskiness is sometimes noted during infections or crying. Prominence of the left precordium is common, as are
a palpable parasternal lift, a laterally displaced apical impulse and apical thrust, and a systolic thrill. The holosystolic murmur of a large VSD is generally less harsh than that of a small VSD and more blowing in nature because of the absence of a significant pressure gradient across the defect. The pulmonic component of the 2nd heart sound may be increased as a result of pulmonary hypertension.

Diagnosis. In patients with small VSDs, the *chest radiograph* is usually normal, although minimal cardiomegaly and a borderline increase in pulmonary vasculature may be observed. The *ECG* is generally normal but may suggest left ventricular hypertrophy. The presence of right ventricular hypertrophy is a warning that the defect is not small and that the patient has pulmonary hypertension or an associated lesion such as pulmonic stenosis. In large VSDs, the *chest radiograph* shows gross cardiomegaly with prominence of both ventricles, the left atrium, and the pulmonary artery. Pulmonary vascular markings are increased, and frank pulmonary edema, including pleural effusions, may be present. The *ECG* shows biventricular hypertrophy; P waves may be notched or peaked.

The *EchoCG* shows the position and size of the VSD. In small defects, especially those of the muscular septum, the defect itself may be difficult to image and is visualized only by color Doppler examination. In defects of the membranous septum, a thin membrane can partially cover the defect and limit the volume of the left-to-right shunt. Doppler examination shows whether the VSD is pressure restrictive by calculating the pressure gradient across the defect. Such calculation allows an estimation of right ventricular pressure and helps determine whether the patient is at risk for the development of early pulmonary vascular disease.

Catheterization is usually performed only when the size of the shunt is uncertain after a comprehensive clinical evaluation, when laboratory data do not fit well with the clinical findings, or when pulmonary vascular disease is suspected.

Prognosis and Complications. The natural course of a VSD depends on a large degree of the size of the defect. A significant number (30–50 %) of small defects close spontaneously, most frequently during the 1st 2 yr of life. Small muscular VSDs are more likely to close (up to 80 %) than membranous VSDs are (up to 35 %). The vast majority of defects that close do so before the age of 4 yr, although spontaneous closure has been reported in adults. These VSDs often have ventricular septal aneurysms limiting the magnitude of the shunt. Most children with small defects remain asymptomatic, without evidence of an increase in heart size, pulmonary arterial pressure, or resistance. A long-term risk are infective endocarditis, arrhythmia, subaortic stenosis, exercise intolerance. Small, hemodynamically insignificant VSD is not an indication for surgery. All VSDs are closed electively by mid-childhood.

Infants with large defects have repeated episodes of respiratory infection and heart failure despite optimal medical management. Heart failure may be manifested in many of these infants primarily as failure to thrive. Pulmonary hypertension occurs as a result of high pulmonary blood flow. These patients are at risk for pulmonary vascular disease with time if the defect is not repaired.

Patients with VSD are also at risk for the development of aortic valve regurgitation, the greatest risk occurring in patients with supracristal VSD. A small number of patients an Eisenmenger physiology develops.

Treatment. In patients with small VSDs, parents should be reassured of the relatively benign nature of the lesion, and the child should be encouraged to live a normal life, with no restrictions on physical activity. Surgical repair is currently not recommended. As protection against infective endocarditis, the integrity of primary and permanent teeth should be carefully maintained; antibiotic prophylaxis should be provided for dental visits (including cleanings), tonsillectomy, adenoidectomy, and other oropharyngeal surgical procedures,
as well as for instrumentation of the genitourinary and lower intestinal tracts. The ECG is an excellent means of screening these patients for possible pulmonary hypertension or pulmonic stenosis as indicated by right ventricular hypertrophy. EchoCG is used to screen for the development of left ventricular outflow tract pathology (subaortic membrane or aortic regurgitation) and to confirm spontaneous closure.

In infants with a large VSD, medical management has two aims: to control heart failure and prevent the development of pulmonary vascular disease. If early treatment is successful, the shunt may diminish in size with spontaneous improvement, especially during the 1st yr of life. The clinician must be alert to not confuse clinical improvement caused by a decrease in defect size with clinical changes caused by the development of Eisenmenger physiology. Pulmonary vascular disease can be prevented when surgery is performed within the 1st yr of life.

Indications for surgical closure of a VSD include patients at any age with large defects in whom clinical symptoms and failure to thrive cannot be controlled medically; infants between 6 and 12 mo of age with large defects associated with pulmonary hypertension, even if the symptoms are controlled by medication; and patients older than 24 mo with a Qp:Qs ratio greater than 2:1. Severe pulmonary vascular disease is a contraindication to closure of a VSD.

The long-term prognosis after surgery is excellent. Most infants begin to thrive, and cardiac medications are no longer required. Catch-up growth occurs in most patients over the next 1–2 yr.

COARCTATION OF THE AORTA

Constrictions of the aorta of varying degrees may occur at any point from the transverse arch to the iliac bifurcation, but 98 % occur just below the origin of the left subclavian artery at the origin of the ductus arteriosus (juxtaductal coarctation). The anomaly occurs twice as often in males as in females. Coarctation of the aorta may be a feature of Turner syndrome and is associated with a bicuspid aortic valve in more than 70 % of patients.

Pathophysiology. Coarctation of the aorta can occur as a discrete juxtaductal obstruction or as tubular hypoplasia of the transverse aorta starting at one of the head or neck vessels and extending to the ductal area (preductal or infantile-type coarctation). Often, both components are present. It is postulated that coarctation may be initiated in fetal life by the presence of a cardiac abnormality that results in decreased blood flow anterograde through the aortic valve (e. g., bicuspid aortic valve, VSD).

In patients with discrete juxtaductal coarctation, ascending aortic blood flows through the narrowed segment to reach the descending aorta, although left ventricular hypertension and hypertrophy result. In the 1st few days of life, the PDA may serve to widen the juxtaductal area of the aorta and provide temporary relief from the obstruction. Net left-to-right ductal shunting occurs in these acyanotic infants. With more severe juxtaductal coarctation or in the presence of transverse arch hypoplasia, right ventricular blood is ejected through the ductus to supply the descending aorta. Perfusion of the lower part of the body is then dependent on right ventricular output. In this situation, the femoral pulses are palpable, and differential blood pressures may not be helpful in making the diagnosis. The ductal right-to-left shunting is manifested as differential cyanosis, with the upper extremities being pink and the lower extremities blue.

Such infants may have severe pulmonary hypertension and high pulmonary vascular resistance. Signs of heart failure are prominent. Coarctation associated with arch hypoplasia was referred to as *infantile type* because its severity usually led to recognition of the condition in early infancy. *Adult type* referred to isolated juxtaductal coarctation, which if mild, was not usually recognized until later childhood. These terms have been replaced with

the more accurate anatomic terms describing the location and severity of the defect.

Blood pressure is elevated in the vessels that arise proximal to the coarctation; blood pressure as well as pulse pressure is lower below the constriction. Unless operated on in infancy, coarctation of the aorta usually results in the development of an extensive collateral circulation, chiefly from branches of the subclavian, superior intercostal, and internal mammary arteries, to create channels for arterial blood to bypass the area of coarctation. The vessels contributing to the collateral circulation may become markedly enlarged and tortuous by early adulthood.

Clinical Manifestations. Coarctation of the aorta recognized after infancy is rarely associated with significant symptoms. Some children or adolescents complain about weakness or pain (or both) in the legs after exercise, but in many instances, even patients with severe coarctation are asymptomatic. Older children are frequently brought to the cardiologist's attention when they are found to be hypertensive on routine physical examination.

The classic sign of coarctation of the aorta is a disparity in pulsation and blood pressure in the arms and legs. The femoral, popliteal, posterior tibial, and dorsalis pedis pulses are weak (or absent in up to 40 % of patients), in contrast to the bounding pulses of the arms and carotid vessels. The radial and femoral pulses should always be palpated simultaneously for the presence of a radial-femoral delay. Normally, the femoral pulse occurs slightly before the radial pulse.

A radial-femoral delay occurs when blood flow to the descending aorta is dependent on collaterals, in which case the femoral pulse is felt after the radial pulse. In normal persons, systolic blood pressure in the legs obtained by the cuff method is 10–20 mm Hg higher than that in the arms. In coarctation of the aorta, blood pressure in the legs is lower than that in the arms; frequently, it is difficult to obtain. It is important to determine the blood pressure in each arm; a pressure higher in the right than the left arm suggests involvement of the left subclavian artery in the area of coarctation. Occasionally, the right subclavian may arise anomalously from below the area of coarctation and result in a left arm pressure that is higher than the right. With exercise, a more prominent rise in systemic blood pressure occurs, and the upper-to-lower extremity pressure gradient will increase.

The precordial impulse and heart sounds are usually normal; the presence of a systolic ejection click or thrill in the suprasternal notch suggests a bicuspid aortic valve (present in 70 % of cases). A short systolic murmur is often heard along
the left sternal border at the 3rd and 4th intercostal spaces. The murmur is well transmitted to the left infrascapular area and occasionally to the neck. In older patients with well-developed collateral blood flow, systolic or continuous murmurs may be heard over the left and right sides of the chest laterally and posteriorly, a palpable thrill can occasionally be appreciated in the intercostal spaces on the back.

Neonates or infants with more severe coarctation, usually including some degree of transverse arch hypoplasia, initially have signs of lower body hypoperfusion, acidosis, and severe heart failure. These signs may be delayed days or weeks until after closure of the ductus arteriosus. If detected before ductal closure, patients may exhibit differential cyanosis, best demonstrated by simultaneous oximetry of the upper and lower extremities. On physical examination, the heart is large, and a systolic murmur is heard along the left sternal border with a loud 2nd heart sound.

Diagnosis. *X-ray* depends on the age of the patient and on the effects of hypertension and the collateral circulation. Cardiac enlargement and pulmonary congestion are noted in infants with severe coarctation. During childhood, the findings are not striking until after the 1st decade, when the heart tends to be mildly or moderately enlarged because of left ventricular prominence. The enlarged left subclavian artery commonly produces a prominent shadow in the left superior mediastinum. Notching of the inferior border of the ribs from pressure erosion by enlarged collateral vessels is common by late childhood. In most instances, the descending aorta has an area of poststenotic dilatation.

ECG is usually normal in young children but reveals evidence of left ventricular hypertrophy in older patients. Neonates and young infants display right or biventricular hypertrophy. The segment of coarctation can generally be visualized by *EchoCG*; associated anomalies of the mitral and aortic valve can also be demonstrated. The descending aorta is hypopulsatile. Color Doppler is useful for demonstrating the specific site of the obstruction. Pulsed and continuous wave Doppler studies determine the pressure gradient directly at the area of coarctation; in the presence of a PDA, however, the severity of the

narrowing may be underestimated. *Cardiac catheterization* with selective left ventriculography and aortography is useful in certain patients with additional anomalies and as a means of visualizing collateral blood flow. In cases that are well defined by echocardiography, diagnostic catheterization is not usually required before surgery.

Prognosis and Complications. Abnormalities of the aortic valve are present in most patients. Bicuspid aortic valves are common but do not generally produce clinical signs unless the stenosis is significant. The association of a PDA and coarctation of the aorta is also common. VSDs and ASDs may be suspected by signs of a left-to-right shunt; they are exacerbated by the increased resistance to flow through the left side of the heart. Mitral valve abnormalities are also occasionally seen, as is subvalvular aortic stenosis.

Severe neurologic damage or even death may rarely occur from associated cerebrovascular disease. Subarachnoid or intracerebral hemorrhage may result from rupture of congenital aneurysms in the circle of Willis, rupture of other vessels with defective elastic and medial tissue, or rupture of normal vessels; these accidents are secondary to hypertension.

Untreated, the great majority of older patients with coarctation of the aorta would succumb between the ages of 20 and 40 yr; some live well into middle life without serious disability. The common serious complications are related to systemic hypertension, which may result in premature coronary artery disease, heart failure, hypertensive encephalopathy, or intracranial hemorrhage. Heart failure may be worsened by associated anomalies. Infective endocarditis or endarteritis is a significant complication in adults. Aneurysms of the descending aorta or the enlarged collateral vessels may develop. In infants with severe coarctation, heart failure and hypoperfusion may be life threatening and require immediate medical intervention.

Treatment. In neonates with severe coarctation of the aorta, closure of the ductus often results in hypoperfusion, acidosis, and rapid deterioration. These patients should be given an infusion of prostaglandin E_1 to reopen the ductus and re-establish adequate lower extremity blood flow. Once a diagnosis has been confirmed and the patient stabilized, surgical repair should be performed. Older infants with heart failure but good perfusion should be managed with anticongestive measures to improve their clinical status before surgical intervention.

Older children with significant coarctation of the aorta should be treated relatively soon after diagnosis. Delay is unwarranted, especially after the 2nd decade of life, when the operation may be less successful because of decreased left ventricular function and degenerative changes in the aortic wall. Associated valvular lesions increase the hazards of late surgery.

The procedure of choice for isolated juxtaductal coarctation of the aorta is controversial. Surgery remains the treatment of choice, and several surgical techniques are used. The area of coarctation can be excised and a primary re-anastomosis performed. Often, the transverse aorta is splayed open and an “extended end-to-end” anastomosis performed to increase the effective cross-sectional area of the repair. The subclavian flap procedure, which involves division of the left subclavian artery and incorporation of it into the wall of the repaired coarctation, is used by some, often in the younger age group. Others favor a patch aortoplasty, in which the area of coarctation is enlarged with a roof of prosthetic material. The use of angioplasty for native coarctation remains controversial.

After surgery, a striking increase in the amplitude of pulsations in the lower extremities is noted. In the immediate postoperative course, “rebound” hypertension is common and requires medical management. This exaggerated acute hypertension gradually subsides, and in most patients, antihypertensive medications can be discontinued. Residual murmurs are common and may be due to associated cardiac anomalies, to a residual flow disturbance across the repaired area, or to collateral blood flow. Rare operative problems include spinal cord injury from aortic cross-clamping if the collaterals are poorly developed, chylothorax, diaphragm injury, and laryngeal nerve injury. If a left subclavian flap is used, the radial pulse and blood pressure in the left arm are diminished or absent. Repair of coarctation in the 2nd decade of life or beyond may be associated with a higher incidence of premature cardiovascular disease, even in the absence of residual cardiac abnormalities. Early onset of adult chronic hypertension may occur, even in patients with adequately resected coarctation.

Although re-stenosis in older patients after coarctectomy is rare, a significant number of infants operated on before 1 yr of age require revision later in childhood. All patients should be monitored carefully for the development of recoarctation and aortic aneurysm. Should recoarctation occur,

balloon angioplasty is the procedure of choice. In these patients, scar tissue from previous surgery makes reoperation more difficult, yet makes balloon angioplasty safer because of the lower incidence of aneurysm formation. Relief of obstruction with this technique is usually excellent. Intravascular stents are now commonly used in many of these patients with generally excellent results.

TETRALOGY OF FALLO

The tetralogy of Fallot consists of: 1) obstruction to right ventricular outflow (pulmonary stenosis), 2) VSD, 3) dextroposition of the aorta with septal override, 4) right ventricular hypertrophy.

***Pathophysiology*.** The pulmonary valve annulus may be of nearly normal size or quite small. The valve itself is often bicuspid and, occasionally, is the only site of stenosis. More commonly, the subpulmonic muscle, the crista supraventricularis, is hypertrophic, which contributes to the infundibular stenosis and results in an infundibular chamber of variable size and contour. When
the right ventricular outflow tract is completely obstructed (pulmonary atresia), the anatomy of the branch pulmonary arteries is extremely variable; a main pulmonary artery segment may be in continuity with right ventricular outflow, separated by a fibrous but imperforate pulmonary valve, or the entire main pulmonary artery segment may be absent. Occasionally, the branch pulmonary arteries may be discontinuous. In these more severe cases, pulmonary blood flow may be supplied by a patent ductus arteriosus and by *major aortopulmonary collateral arteries (MAPCAs)* arising from the aorta.

The VSD is usually nonrestrictive and large, is located just below the aortic valve, and is related to the posterior and right aortic cusps.

Systemic venous return to the right atrium and right ventricle is normal. When the right ventricle contracts in the presence of marked pulmonary stenosis, blood is shunted across the VSD into the aorta. Persistent arterial desaturation and cyanosis result. Pulmonary blood flow, when severely restricted by the obstruction to right ventricular outflow, may be supplemented by the bronchial collateral circulation (MAPCAs) and, in the newborn, by a PDA. Peak systolic and diastolic pressures in each ventricle are similar as at the systemic level. A large pressure gradient occurs across the obstructed right ventricular outflow tract, and pulmonary arterial pressure is normal or lower

than normal. The degree of right ventricular outflow obstruction determines the timing of the onset of symptoms, the severity of cyanosis, and the degree of right ventricular hypertrophy. When obstruction to right ventricular outflow is mild to moderate and a balanced shunt is present across the VSD, the patient may not be visibly cyanotic *(acyanotic or "pink" tetralogy of Fallot)*.

***Clinical Manifestations*.** Infants with mild degrees of right ventricular outflow obstruction may initially be seen with heart failure caused by a ventricular-level left-to-right shunt. Often, cyanosis is not present at birth, but with increasing hypertrophy of the right ventricular infundibulum and patient growth, cyanosis occurs later in the 1st yr of life. It is most prominent in the mucous membranes of the lips and mouth and in the fingernails and toenails. In infants with severe degrees of right ventricular outflow obstruction, neonatal cyanosis is noted immediately. In these infants, pulmonary blood flow may be dependent on flow through the ductus arteriosus. When the ductus begins to close in the 1st few hours or days of life, severe cyanosis and circulatory collapse may occur. Older children with long-standing cyanosis who have not undergone surgery may have dusky blue skin, gray sclerae with engorged blood vessels, and marked *clubbing* of the fingers and toes.

Dyspnea occurs on exertion. Infants and toddlers play actively for a short time and then sit or lie down. Older children may be able to walk a block or so before stopping to rest. Characteristically, children assume a *squatting* position for the relief of dyspnea caused by physical effort; the child is usually able to resume physical activity within a few minutes. These findings occur most often in patients with significant cyanosis at rest.

Paroxysmal hypercyanotic attacks (hypoxic, "blue" or "tet" spells) are a particular problem during the 1st 2 yr of life. The infant becomes hyperpneic and restless, cyanosis increases, gasping respirations ensue, and syncope may follow. The spells occur most frequently in the morning on initially awakening or after episodes of vigorous crying. Temporary disappearance or a decrease in intensity of the systolic murmur is usual as flow across the right ventricular outflow tract diminishes. The spells may last from a few minutes to a few hours but are rarely fatal. Short episodes are followed by generalized weakness and sleep. Severe spells may progress to unconsciousness and, occasionally, to convulsions or hemiparesis. The onset is usually spontaneous and unpredictable. Spells are associated with reduction of an already compromised pulmonary

blood flow, which when prolonged results in severe systemic hypoxia and metabolic acidosis. Infants who are only mildly cyanotic at rest are often more prone to the development of hypoxic spells because they have not acquired the homeostatic mechanisms to tolerate rapid lowering of arterial oxygen saturation, such as polycythemia.

Immediate care:

1) placement of the infant on the abdomen in the knee-chest position while making certain that the infant's clothing is not constrictive;

2) administration of oxygen;

3) injection of morphine subcutaneously in a dose not in excess of 0.2 mg/kg. Calming and holding the infant in a knee-chest position may abort progression of an early spell;

4) rapid correction of metabolic acidosis with intravenous $NaHCO_3$;

5) β-adrenergic blockade — propranolol (0.1 mg/kg given slowly to a maximum of 0.2 mg/kg) IV is also useful.

Growth and development may be delayed in patients with severe untreated tetralogy of Fallot, particularly when oxygen saturation is chronically less than 70 %. Puberty may also be delayed in patients who do not undergo surgery.

The pulse is usually normal, as is venous and arterial pressure. The left anterior hemithorax may bulge anteriorly because of right ventricular hypertrophy. The heart is generally normal in size, and a *substernal right ventricular impulse* can be detected. In about half the cases, a *systolic thrill* is felt along the left sternal border in the 3rd and 4th parasternal spaces. The *systolic murmur* is usually loud and harsh; it may be transmitted widely, especially to the lungs, but is most intense at the left sternal border. The murmur is generally ejection in quality at the upper sternal border, but it may sound more holosystolic toward the lower sternal border. It may be preceded by a click. The murmur is caused by turbulence through the right ventricular outflow tract. It tends to become louder, longer, and harsher as the severity of pulmonary stenosis increases from mild to moderate; however, it can actually become less prominent with severe obstruction, especially during a hypercyanotic spell. Either the 2nd heart sound

is single, or the pulmonic component is soft. Infrequently, a continuous murmur may be audible, especially if prominent collaterals are present.

***Diagnosis*.** The typical configuration as seen on the *X-ray*: anteroposterior view consists of a narrow base, concavity of the left heart border in the area usually occupied by the pulmonary artery, and normal heart size. The hypertrophied right ventricle causes the rounded apical shadow to be up-tilted so that it is situated higher above the diaphragm than normal. The cardiac silhouette has been likened to that of a boot or wooden shoe (*coeur en sabot*). The hilar areas and lung fields are relatively clear because of diminished pulmonary blood flow or the small size of the pulmonary arteries, or both. The aorta is usually large, and in about 20 % of instances it arches to the right, which results in an indentation of the leftward-positioned air-filled tracheobronchial shadow in the anteroposterior view.

ECG right axis deviation and evidence of right ventricular hypertrophy. A dominant R wave appears in the right precordial chest leads (Rs, R, qR, qRs). In some cases, the only sign of right ventricular hypertrophy may initially be a positive T wave in leads V_3R and V_1. The P wave is tall and peaked or sometimes bifid.

EchoCG establishes the diagnosis and provides information about the extent of aortic override of the septum, the location and degree of the right ventricular outflow tract obstruction, the size of the proximal branch pulmonary arteries, and the side of the aortic arch.

Cardiac catheterization demonstrates a systolic pressure in the right ventricle equal to systemic pressure. If the pulmonary artery is entered, the pressure is markedly decreased, although crossing the right ventricular outflow tract, especially in severe cases. Pulmonary arterial pressure is usually lower than normal, in the range of 5–10 mm Hg. The level of arterial oxygen saturation depends on the magnitude of the right-to-left shunt; in “pink tets”, systemic saturation may be normal, whereas in a moderately cyanotic patient at rest, it is usually 75–85 %.

Complete and accurate information regarding the anatomy of the vessels (*selective right ventriculography, left ventriculography, aortography* or *coronary arteriography)* is important when evaluating these children as surgical candidates.

Prognosis and Complications. Before correction, patients with the tetralogy of Fallot are susceptible to several serious complications. Fortunately, most children undergo palliation or repair in infancy, and these complications are rare. *Cerebral thromboses*, usually occurring in the cerebral veins or dural sinuses and occasionally in the cerebral arteries, are common in the presence of extreme polycythemia and dehydration. Thromboses occur most often in patients younger than 2 yr. These patients may have iron deficiency anemia, frequently with hemoglobin and hematocrit levels in the normal range. Therapy consists of adequate hydration and supportive measures. Phlebotomy and volume replacement with fresh frozen plasma are indicated in extremely polycythemic patients. Heparin is of little value and is contraindicated in patients with hemorrhagic cerebral infarction. Physical therapy should be instituted as early as possible.

Brain abscess is less common than cerebral vascular events and extremely rare when most patients are repaired at much younger ages. Patients with a brain abscess are usually older than 2 yr. The onset of the illness is often insidious and consists of low-grade fever or a gradual change in behavior, or both. Some patients have an acute onset of symptoms that may develop after a recent history of headache, nausea, and vomiting. Seizures may occur; localized neurologic signs depend on the site and size of the abscess and the presence of increased intracranial pressure. CT or MRI confirms the diagnosis. Antibiotic therapy may help keep the infection localized, but surgical drainage of the abscess is usually necessary.

Bacterial endocarditis may occur in the right ventricular infundibulum or on the pulmonic, aortic, or rarely, the tricuspid valves. Endocarditis may complicate palliative shunts or, in patients with corrective surgery, any residual pulmonic stenosis or VSD. Antibiotic prophylaxis is essential before and after dental and certain surgical procedures associated with a high incidence of bacteremia.

Heart failure is not a usual feature in patients with the tetralogy of Fallot. It may occur in a young infant with “pink” or acyanotic tetralogy of Fallot. As the degree of pulmonary obstruction worsens with age, the symptoms of heart failure resolve and eventually the patient experiences cyanosis, often by 6–12 mo of age. These patients are at increased risk for hypercyanotic spells at this time.

Treatment. Depends on the severity of the right ventricular outflow tract obstruction. Infants with severe tetralogy require medical treatment and surgical intervention in the neonatal period. Therapy is aimed at providing an immediate increase in pulmonary blood flow to prevent the sequelae of severe hypoxia.

The infant should be transported to a medical center adequately equipped to evaluate and treat neonates with CHD. It is critical that oxygenation and normal body temperature be maintained during the transfer. Prolonged, severe hypoxia may lead to shock, respiratory failure, and intractable acidosis and will significantly reduce the chance of survival, even when surgically amenable lesions are present. Blood glucose levels should be monitored because hypoglycemia is more likely to develop in infants with cyanotic heart disease.

Infants with marked right ventricular outflow tract obstruction may deteriorate rapidly because as the ductus arteriosus begins to close, pulmonary blood flow is further compromised. The IV administration of prostaglandin E_1 (0.05–0.20 mg/kg/min), a potent and specific relaxant of ductal smooth muscle, causes dilatation of the ductus arteriosus and usually provides adequate pulmonary blood flow until a surgical procedure can be performed. Should be administered IV as soon as cyanotic CHD is clinically suspected and continued through

the preoperative period and during cardiac catheterization. Postoperatively, the infusion may be continued briefly as a pulmonary vasodilator to augment flow through a palliative shunt or through a surgical valvulotomy.

Infants with less severe right ventricular outflow tract obstruction who are stable and awaiting surgical intervention require careful observation. Prevention or prompt treatment of dehydration is important to avoid hemoconcentration and possible thrombotic episodes. Paroxysmal dyspneic attacks in infancy or early childhood may be precipitated by a relative iron deficiency; iron therapy may decrease their frequency and also improve exercise tolerance and general well-being. Red blood cell indices should be maintained in the normocytic range. Oral propranolol (0.5–1 mg/kg every 6 hr) may decrease the frequency and severity of hypercyanotic spells, but with the excellent surgery available, surgical treatment is indicated as soon as spells begin.

The modified *Blalock–Taussig shunt* is currently the most common aortopulmonary shunt procedure and consists of a Gore-Tex conduit anastomosed side to side from the subclavian artery to the homolateral branch of the pulmonary artery. Sometimes the conduit is brought directly from the ascending aorta to the main pulmonary artery and is called a *central shunt*. The Blalock–Taussig operation can be successfully performed in the newborn period with shunts 3–4 mm in diameter and has also been used successfully in premature infants.

After a successful shunt procedure, cyanosis diminishes. The development of a continuous murmur over the lung fields after the operation indicates a functioning anastomosis. A good shunt murmur may not be heard until several days after surgery. The duration of symptomatic relief is variable. As the child grows, more pulmonary blood flow is needed and the shunt eventually becomes inadequate. When increasing cyanosis develops, a corrective operation should be performed if the anatomy is favorable. If not possible (e. g., because of hypoplastic branch pulmonary arteries) or if the 1st shunt lasts only a brief period in a small infant, a second aortopulmonary anastomosis may be required on the opposite side. Several groups have reported successful palliation of the tetralogy of Fallot in infants by balloon pulmonary valvuloplasty.

Corrective surgical therapy consists of relief of the right ventricular outflow tract obstruction by removing obstructive muscle bundles and patch closure of the VSD. The surgical risk of total correction is less than 5 %. A right ventriculotomy was the standard approach; however, a transatrial-transpulmonary approach can be used to reduce the long-term risks of a ventriculotomy.

After successful total correction, patients are generally asymptomatic and are able to lead unrestricted lives. Immediate postoperative problems include right ventricular failure, transient heart block, residual VSD with left-to-right shunting, myocardial infarction from interruption of an aberrant coronary artery, and disproportionately increased left atrial pressure because of residual bronchial collaterals. Postoperative heart failure (particularly in patients with a transannular outflow patch) requires a positive inotropic agent such as digoxin. The majority of patients after tetralogy repair and all of those with transannular

patch repairs have a to-and-fro murmur at the left sternal border, usually indicative of mild outflow obstruction and mild to moderate pulmonary insufficiency. Patients with more marked pulmonary valve insufficiency also have moderate to marked heart enlargement. Patients with a severe residual gradient across the right ventricular outflow tract may require reoperation, but mild to moderate obstruction is virtually always present and does not require re-intervention.

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