Chapter 1
Diagnostic methods

The history and physical examination are the keystones for diagnosis of cardiac problems. A variety of other diagnostic techniques can be employed beyond the history and physical examination. With each technique, different aspects of the cardiovascular system are viewed, and by combining the data derived, an accurate assessment of the patient's condition can be obtained.

HISTORY

General principles of the cardiovascular history
The suspicion of a cardiovascular abnormality may be raised initially by specific symptoms, but more commonly the presenting feature is the discovery of a cardiac murmur. Many children with a cardiac abnormality are asymptomatic

2 Pediatric cardiology

because the malformation does not result in major hemodynamic alterations. Even with a significant cardiac problem, the child may be asymptomatic because the myocardium is capable of responding normally to the stresses placed upon it by the altered hemodynamics. A comparable lesion in an adult might produce symptoms because of coexistent coronary arterial disease or myocardial fibrosis.

In obtaining the history of a child suspected of cardiac disease, the physician seeks three types of data.

Diagnostic clues

Diagnostic clues (e.g., cyanosis or squatting) and other more general factors include the following.

Gender. Certain cardiac malformations have a definite gender predominance. Atrial septal defect (ASD) and patent ductus arteriosus are two to three times more likely in female than in male children. Coarctation of the aorta, aortic stenosis, and transposition of the great vessels more commonly occur in male children.

Age. The age at which a cardiac murmur or a symptom develops may give a diagnostic clue. The murmurs of congenital aortic stenosis and pulmonary stenosis are often heard in the neonatal period. Ventricular septal defect (VSD) is usually first recognized because of symptoms and murmur, at 4–6 weeks. The murmur of an ASD may not be discovered until the preschool examination. Among school age children, a functional (innocent) murmur is found in half.

Severity of the cardiac condition

The physician seeks information that suggests the condition's severity (e.g., dyspnea).

Etiology

The physician seeks information that suggests an etiology of cardiac disease (e.g., maternal lupus).

Chief complaint and/or presenting sign

Certain presenting complaints and signs are more common in certain cardiac disorders and the "index of suspicion" aids the physician in organizing the data to make a differential diagnosis. For many of the signs and symptoms discussed later, *noncardiac causes are often more likely than cardiac causes* (e.g., acute dyspnea in a previously healthy 4-month-old infant with no murmur is more likely a result of bronchiolitis than of congestive heart failure). Therefore, a complete history must be integrated with the physical examination and other diagnostic studies to arrive at the correct cardiac diagnosis.

Murmur

Murmur is the most common presenting finding because virtually all children and adults with a normal heart have an innocent (normal) murmur heard sometime during their lifetime. Certain historical features are associated with an innocent murmur; the child is asymptomatic and murmurs appearing after infancy tend to be innocent. The murmur of atrial septal defect is one important exception.

Chest pain

Chest pain is a common and benign symptom in older children and adolescents, estimated to occur at some time in 70% of school-aged children. About 1 in 200 visits to a pediatric emergency room is for chest pain.

   Chest pain rarely occurs with cardiovascular disease in this age group. Myocardial ischemic syndromes (e.g., Kawasaki disease with aneurysms; hypertrophic cardiomyopathy) may lead to true angina. Patients with connective tissue disorders (e.g., Marfan syndrome) may have chest (or back) pain from arterial dissection. Although pericarditis may cause chest pain, it is almost always associated with fever and other signs of inflammation. Occasionally, chest pain accompanies supraventricular tachycardia. The vast majority of children with congenital cardiac malformations, including those who are fully recovered from surgery, do not have chest pain, and the majority of children and adolescents who do present with chest pain as their chief complaint do not have cardiac malformations or disease.

   Benign chest pain is usually transient, appearing abruptly, lasting 30 seconds to 5 minutes and localized to the parasternal area. It can be distinguished from angina by the absence of diaphoresis, nausea, emesis, and paresthesias in an ulnar distribution. Benign chest pain is "sharp," not "crushing" like angina. Chest pain may occur as a result of chest wall tenderness, which is typically well localized, sharp in character, short (seconds to minutes) in duration, often aggravated by certain positions or movements, and can be induced by palpation over the area. These findings tend to be good evidence against cardiac cause for the pain. Some other noncardiac conditions (e.g., asthma) may be associated with childhood chest pain. However, benign pain is often described as "functional" because an organic cause cannot be found.

Palpitations

Palpitations, the sensation of irregular heartbeats, "skipped beats," or more commonly, rapid beats, are also common in the school-aged child and adolescent. They frequently occur in patients with other symptoms, such as chest pain, but often not simultaneously with the other symptoms. Palpitations are often found to be associated with normal sinus rhythm when the electrocardiogram is monitored during the symptom. Palpitations are not usually present in patients

with known premature beats. Palpitations of sudden onset (approximately the time span of a single beat) and termination suggest tachyarrhythmia.

Near-syncope

Near-syncope is a complex of symptoms that includes vertigo and weakness. It is often induced by a postural change (orthostatic), is found commonly in older children and adolescents, and is almost always benign. The history often reveals little fluid and caloric intake beforehand. *True syncope*, characterized by complete loss of consciousness and loss of skeletal muscle tone, rarely results from a cardiac abnormality. It is often autonomic (vasovagal) in origin. Benign syncope is usually very brief in duration, often lasting only seconds. Benign syncope may follow a period of physical activity by several minutes; however, *syncope during exercise* often indicates a serious cardiac problem, such as aortic stenosis, arrhythmia, or myocardial abnormality. Because some life-threatening conditions (e.g., long QT syndrome) may result in syncope after a patient has been startled or has experienced an emotionally stressful situation, similar to benign syncope, an electrocardiogram is advisable for any child with a history of syncope. The family history should be explored for sudden death, syncope, seizures, SIDS, swimming deaths, and single-occupant motor vehicle fatalities.

Dyspnea

Dyspnea (labored breathing) is a symptom present in patients with pulmonary congestion from either left-sided cardiac failure or other conditions raising pulmonary venous pressure or from marked hypoxia. Dyspnea is manifested in infants by rapid, grunting respirations associated with retractions. Older children complain of shortness of breath. The most common causes in children are asthma and bronchitis.

Fatigue

Fatigue on exercise must be distinguished from dyspnea as it has a different physiologic basis.

> Exercise intolerance of cardiac origin indicates an inability of the heart to meet the increased metabolic demands for oxygen delivery to the tissues during this state. This can occur in three situations:
> *Cyanotic congenital heart disease* (arterial oxygen desaturation).
> *Congestive cardiac failure* (inadequate myocardial function).
> *Severe outflow obstructive conditions or those causing cardiac filling impairment* (inadequate cardiac output).

Fatigue on exercise or exercise intolerance is a difficult symptom to interpret because other factors, such as motivation or amount of training, influence the amount of exercise an individual can perform. To assess exercise intolerance, compare the child's response to physical activity with that of peers and siblings or to their previous level of activity.

Growth retardation
Growth retardation is common in many children who present with other cardiac symptoms within the first year of life.

Infants with cardiac failure or cyanosis. Infants with cardiac failure or cyanosis show retarded growth, which is more marked if they both are present. Usually, the rate of weight increase is more delayed than that of height. The cause of growth retardation is unknown, but it is probably related to inadequate caloric intake due to dyspnea during feeding and to the excessive energy requirements of congestive cardiac failure.

Growth. Growth may also be retarded in children with a cardiac anomaly associated with a syndrome, such as Down syndrome, which in itself causes growth retardation.

Developmental milestones. Developmental milestones requiring muscle strength may be delayed, but usually mental development is normal. In assessing growth and development, obtaining growth development data for siblings as well as for the parents and grandparents is helpful.

Congestive cardiac failure
Congestive cardiac failure leads to the most frequently described symptom complex in infants and children with cardiac disease. In infants and children, 80% of instances of heart failure occur during the first year of life; these are usually associated with a cardiac malformation. The remaining 20% occurring during childhood are usually related to acquired conditions. Infants with cardiac failure are described as slow feeders who tire when feeding, this symptom indicating dyspnea on exertion (the act of sucking a bottle). The infant perspires excessively, presumably from increased catecholamine release. Rapid respiration, particularly when the infant is asleep, is an invaluable clue for cardiac failure in the absence of pulmonary disease. The ultimate diagnosis of cardiac failure rests on a compilation of information from the history, the physical examination, and laboratory studies such as chest X-ray.

Respiratory infections
Respiratory infections, particularly pneumonia, are frequently present in infants and, less commonly, in older children with cardiac anomalies, especially those

6 Pediatric cardiology

associated with increased pulmonary blood flow (left-to-right shunt) or with a greatly enlarged heart. The factors leading to the increased incidence of pneumonia are largely unknown but may be related to compression of the major bronchi by either enlarged pulmonary arteries, an enlarged left atrium, or distended pulmonary lymphatics.

Atelectasis may also occur, particularly in the right upper or middle lobe, in children with greatly increased pulmonary blood flow; or in the left lower lobe in children with cardiomyopathies and massively dilated left hearts.

Cyanosis

Cyanosis is a bluish or purplish color of the skin caused by the presence of at least 5 g/dL of reduced hemoglobin in capillary beds. The desaturated blood imparts a bluish color to the appearance, particularly in areas with a rich capillary network, such as the lips or oral mucosa. The degree of cyanosis reflects the magnitude of unsaturated blood. Mild degrees of arterial desaturation may be present without cyanosis being noted. Usually, if the systemic arterial oxygen saturation is less than 88%, cyanosis can be recognized—this varies with skin pigmentation, adequacy of lighting, and experience of the observer. A minimal degree of cyanosis may appear as a mottled complexion, darkened lips, or plethoric fingertips. Clubbing develops with more significant degrees of cyanosis.

Cyanosis is classified as either peripheral or central.

Peripheral cyanosis. Peripheral cyanosis, also called *acrocyanosis*, is associated with normal cardiac and pulmonary function. Related to sluggish blood flow through capillaries, the continued oxygen extraction eventually leads to increased amounts of desaturated blood in the capillary beds. It typically involves the extremities and usually spares the trunk and mucous membranes. Exposure to cold is the most frequent cause of acrocyanosis, leading to blue hands and feet in neonates and circumoral cyanosis in older children. The cyanosis disappears upon warming. The normal polycythemia of neonates may contribute to the appearance of acrocyanosis.

Central cyanosis. Central cyanosis is related to any abnormality of the lungs, heart, or hemoglobin that interferes with oxygen transport from the atmosphere to systemic capillaries. Cyanosis of this type involves the trunk and mucous membranes as well as the extremities. A variety of pulmonary conditions, such as atelectasis, pneumothorax, and respiratory distress syndrome, can cause cyanosis. Areas of the lungs, though not ventilated, are perfused, and blood flowing through that portion of the lung is unoxygenated. Thus, desaturated blood returns to the left atrium and mixes with fully saturated blood from the ventilated portions of the lungs. Rarely, dysfunctional hemoglobin

disorders, such as excessive levels of methemoglobin, result in cyanosis because hemoglobin is unable to bind normal quantities of oxygen.

Cardiac conditions cause central cyanosis by either of two mechanisms.
(1) *Structural abnormalities.* Structural abnormalities which divert portions of the systemic venous return (desaturated blood) away from the lungs can be caused by two categories of cardiac anomalies:
   (a) *Conditions with obstruction to pulmonary blood flow and an intracardiac septal defect* (e.g., tetralogy of Fallot).
   (b) *Conditions in which the systemic venous and pulmonary venous returns are mixed in a common chamber before being ejected* (e.g., single ventricle).
(2) *Pulmonary edema of cardiac origin*. Mitral stenosis and similar conditions raise pulmonary capillary pressure. When capillary pressure exceeds oncotic pressure, fluid crosses the capillary wall into alveoli. The fluid accumulation interferes with oxygen transport from the alveolus to the capillary so that hemoglobin leaving the capillaries remains desaturated.
   Cyanosis resulting from pulmonary edema may be strikingly improved by oxygen administration, whereas cyanosis occurring with structural cardiovascular anomalies may show little change with this maneuver.

Squatting

Squatting is a relatively specific symptom, occurring almost exclusively in patients with tetralogy of Fallot. When experiencing a hypercyanotic or "tet" spell, cyanotic infants assume a knee/chest position, whereas older children squat in order to rest. In this position the systemic arterial resistance rises, the right-to-left shunt decreases, and the patient becomes less desaturated.

Neurologic symptoms

Neurologic symptoms may occur in children with cardiac disease, particularly those with cyanosis but are seldom the presenting symptoms. Rarely, in otherwise normal children, *seizures* stem from arrhythmias, such as the ventricular tachycardia seen in the long QT syndrome, and are the sole presenting symptom.

Prenatal history

A prenatal history may also suggest an etiology of the cardiac malformation if it yields information, such as maternal rubella, drug ingestion, or other teratogen.

8   Pediatric cardiology

Family history

The physician should obtain a complete family history and pedigree to disclose the presence of congenital cardiac malformations, syndromes, or other disorders, such as hypertrophic cardiomyopathy (associated with sudden death in young persons) or long QT syndrome (associated with a family history of seizures, syncope, and sudden death).

Other historical facts that may be diagnostically significant will be discussed in relation to specific cardiac anomalies.

PHYSICAL EXAMINATION

When examining a child with suspected cardiac abnormalities, the physician may focus too quickly on the auscultatory findings, overlooking the general physical characteristics of the child. In some patients, these findings equal the diagnostic value of the cardiovascular findings.

Cardiac abnormalities are often an integral part of generalized diseases and syndromes: recognition of the syndrome can often provide a clinician with either an answer or a clue as to the nature of the associated cardiac disease. These syndromes are discussed in Chapter 2.

Vital signs
Blood pressure

In all patients suspected of cardiac disease, examiners should record accurately the blood pressure in both arms and one leg. Doing this aids in diagnosis of conditions causing obstruction, such as coarctation of the aorta: recognition of conditions with "aortic runoff," such as patent ductus arteriosus, and identification of reduced cardiac output.

Many errors can be made in obtaining the blood pressure recording. The patient should be in a quiet, resting state, and the extremity in which blood pressure is being recorded should be at the same level as the heart. A properly sized blood pressure cuff must be used because an undersized cuff causes false elevation of the blood pressure reading. A slightly oversized cuff is unlikely to greatly affect readings. Therefore, blood pressure cuffs of various sizes should be available. A guide to the appropriate size for each age group is given in Table 1.1. Generally, the width of the inflatable bladder within the cuff should be at least 40% of the circumference of the limb, and the bladder length should encompass 80–100% of the circumference of the limb at the point of measurement. In infants, placing the cuff around the forearm and leg rather than around the arm and thigh is easier.

Although a 1-inch-wide cuff is available, it should never be used because it leads uniformly to a falsely elevated pressure except in the tiniest prematures. A 2-inch-wide cuff can be used for almost all infants.

Failure to pause between readings does not allow adequate time for return of venous blood trapped during the inflation and may falsely elevate the next reading.

Table 1.1 Recommended Dimensions for Blood Pressure Cuff Bladders.

Age Range	Width (cm)	Length (cm)	Maximum Arm Circumference (cm)*
Newborn	4	8	10
Infant	6	12	15
Child	9	18	22
Small adult	10	24	26
Adult	13	30	34
Large adult	16	38	44
Thigh	20	42	52

*Calculated so that the largest arm would still allow bladder to encircle arm by at least 80%. Adapted from National High Blood Pressure Education Program Working Group on High Blood Pressure in Children and Adolescents. The fourth report on the diagnosis, evaluation, and treatment of high blood pressure in children and adolescents. Pediatrics 2004; 114:555–576.

This is a work of the US government, published in the public domain by the American Academy of Pediatrics, available online at http://pediatrics.aappublications. org/cgi/content/full/114/2/S2/555 and http://www.nhlbi.nih.gov/health/prof/heart/hbp/ hbp_ped.pdf.

Methods. Four methods of obtaining blood pressure can be used in infants and children—three manual methods (flush, palpatory, and auscultatory) and an automated method (oscillometric).

For manual methods, the cuff should be applied snugly and the manometer pressure quickly elevated. The pressure should then be released at a rate of 1–3 mm Hg/s and allowed to fall to zero. After a pause, the cuff can be reinflated. Pressure recordings should be repeated at least once.

Flush method. In this method used in infants, a blood pressure cuff is placed about the infant's extremity, and the hand or foot is tightly squeezed by the physician's hand. The cuff is rapidly inflated, and the infant's hand or foot is released. As the pressure slowly falls, the pressure value at which the blanched hand or foot flushes reflects the mean arterial pressure. In infants, blood pressure should be taken simultaneously in an upper and lower extremity because fussiness and crying lead to falsely elevated, rapidly changing values. The practitioner can accomplish this by connecting two blood pressure cuffs by a Y-tubing to a manometer and placing one cuff on the arm and the other cuff on the leg, thereby obtaining the pressure simultaneously by the flush method.

Palpation. Palpation can also be used in infants. During release of the pressure from the cuff, the pressure reading at which the pulse appears distal to the cuff indicates the systolic blood pressure. A more precise but similar method uses an ultrasonic Doppler probe to register the arterial pulse in lieu of palpating it.
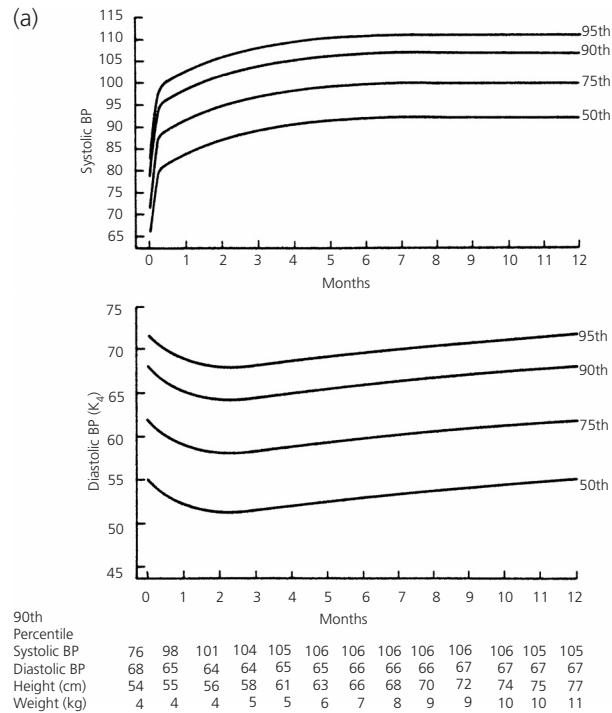
10 Pediatric cardiology

Figure 1.1 Blood pressure upper limits for boys and girls from birth to 1 year of age. (a) Girls; (b) boys. (Adapted from Report of the second task force on blood pressure control in children. Pediatrics 1987;79:1–25.)

Auscultation. In the older child, blood pressure can be obtained by the auscultatory method: in the arm, by listening over the brachial artery in the antecubital space, or in the leg and in the thigh, by listening to the popliteal artery. The pressure at which the first Korotkoff sound (K_1) is heard represents the systolic pressure. As the cuff pressure is released, the pressure at which the sound muffles (K_4) and the pressure at which the sound disappears (K_5) should also be recorded. The diastolic blood pressure is located between these two values.

Automated. Oscillometric methods utilize a machine that automatically inflates and deflates the cuff while monitoring pulse-related air pressure fluctuations within the cuff. Deflation is performed in stepwise fashion, and at each step the machine pauses for 2 seconds or less while the cuff pressure oscillations

90th Percentile												
Systolic BP	76	98	101	104	105	106	106	106	106	106	106	105	105
Diastolic BP	68	65	64	64	65	65	66	66	66	67	67	67	67
Height (cm)	54	55	56	58	61	63	66	68	70	72	74	75	77
Weight (kg)	4	4	4	5	5	6	7	8	9	9	10	10	11

(b)

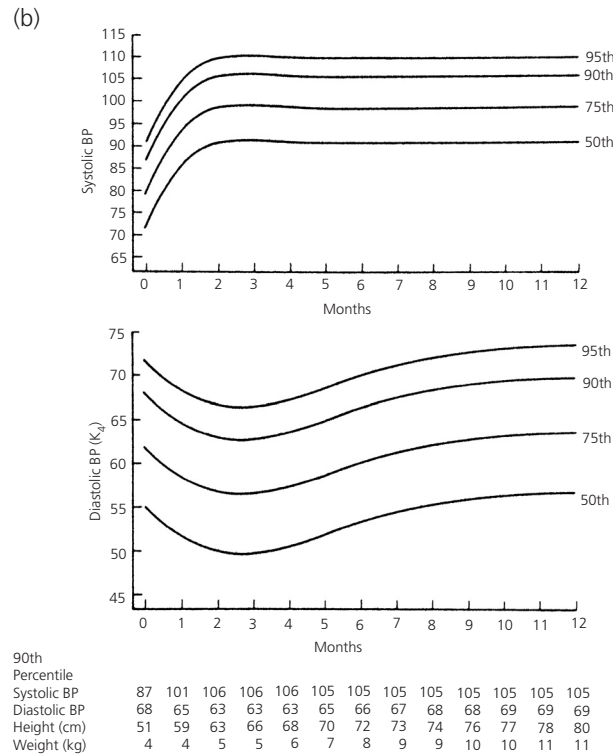| 90th Percentile | | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Systolic BP | 87 | 101 | 106 | 106 | 106 | 105 | 105 | 105 | 105 | 105 | 105 | 105 | 105 |
| Diastolic BP | 68 | 65 | 63 | 63 | 63 | 65 | 66 | 67 | 68 | 68 | 69 | 69 | 69 |
| Height (cm) | 51 | 59 | 63 | 66 | 68 | 70 | 72 | 73 | 74 | 76 | 77 | 78 | 80 |
| Weight (kg) | 4 | 4 | 5 | 5 | 6 | 7 | 8 | 9 | 9 | 10 | 10 | 11 | 11 |

Figure 1.1 (*cont.*)

are recorded. The amplitude of these pulsatile oscillations begins to increase as the cuff pressure falls to the level of the systolic blood pressure, reaches a maximum amplitude at a cuff pressure equal to mean blood pressure, and diminishes as cuff pressure falls to diastolic levels. Because the method depends on measurement of faint pulsatile pressure oscillations, irregular heart rhythm (e.g., atrial fibrillation), conditions with beat-to-beat variability in pulse pressure (e.g., the pulsus alternans of heart failure or mechanical ventilator-induced changes), and patient movement may lead to inaccurate or absent readings.

Normal values. The normal blood pressure values for different age groups are given in Fig. 1.1 and Tables 1.2 and 1.3. The blood pressure in the leg should be the same as that in the arm. Leg blood pressure should also be taken with an

12   Pediatric cardiology

Table 1.2 Blood Pressure Levels for Boys by Age (1–17 years) and Height Percentile.

Age (year)	BP Percentile	Systolic BP (mm Hg) ←Percentile of Height→							Diastolic BP (mm Hg) ←Percentile of Height→						
		5th	10th	25th	50th	75th	90th	95th	5th	10th	25th	50th	75th	90th	95th
1	50th	80	81	83	85	87	88	89	34	35	36	37	38	39	39
	90th	94	95	97	99	100	102	103	49	50	51	52	53	53	54
	95th	98	99	101	103	104	106	106	54	54	55	56	57	58	58
	99th	105	106	108	110	112	113	114	61	62	63	64	65	66	66
2	50th	84	85	87	88	90	92	92	39	40	41	42	43	44	44
	90th	97	99	100	102	04	105	106	54	55	56	57	58	58	59
	95th	101	102	104	106	108	109	110	59	59	60	61	62	63	63
	99th	109	110	111	113	115	117	117	66	67	68	69	70	71	71
3	50th	86	87	89	91	93	94	95	44	44	45	46	47	48	48
	90th	100	101	103	105	107	108	109	59	59	60	61	62	63	63
	95th	104	105	107	109	110	112	113	63	63	64	65	66	67	67
	99th	111	112	114	116	118	119	120	71	71	72	73	74	75	75
4	50th	88	89	91	93	95	96	97	47	48	49	50	51	51	52
	90th	102	103	105	107	109	110	111	62	63	64	65	66	66	67
	95th	106	107	109	111	112	114	115	66	67	68	69	70	71	71
	99th	113	114	116	118	120	121	122	74	75	76	77	78	78	79
5	50th	90	91	93	95	96	98	98	50	51	52	53	54	55	55
	90th	104	105	106	108	110	111	112	65	66	67	68	69	69	70
	95th	108	109	110	112	114	115	116	69	70	71	72	73	74	74
	99th	115	116	118	120	121	123	123	77	78	79	80	81	81	82

Age	Percentile	SBP							DBP						
6	50th	91	92	94	96	98	99	100	53	53	54	55	56	57	57
	90th	105	106	108	110	111	113	113	68	68	69	70	71	72	72
	95th	109	110	112	114	115	117	117	72	72	73	74	75	76	76
	99th	116	117	119	121	123	124	125	80	80	81	82	83	84	84
7	50th	92	94	95	97	99	100	101	55	55	56	57	58	59	59
	90th	106	107	109	111	113	114	115	70	70	71	72	73	74	74
	95th	110	111	113	115	117	118	119	74	74	75	76	77	78	78
	99th	117	118	120	122	124	125	126	82	82	83	84	85	86	86
8	50th	94	95	97	99	100	102	102	56	57	58	59	60	60	61
	90th	107	109	110	112	114	115	116	71	72	72	73	74	75	76
	95th	111	112	114	116	118	119	120	75	76	77	78	79	79	80
	99th	119	120	122	123	125	127	127	83	84	85	86	87	87	88
9	50th	95	96	98	100	102	103	104	57	58	59	60	61	61	62
	90th	109	110	112	114	115	117	118	72	73	74	75	76	76	77
	95th	113	114	116	118	119	121	121	76	77	79	79	80	81	81
	99th	120	121	123	125	127	128	129	84	85	86	87	88	88	89
10	50th	97	98	100	102	103	105	106	58	59	60	61	61	62	63
	90th	111	112	114	115	117	119	119	73	73	74	75	76	77	78
	95th	115	116	117	119	121	122	123	77	78	79	80	81	81	82
	99th	122	123	125	127	128	130	130	85	86	86	88	88	89	90
11	50th	99	100	102	104	105	107	107	58	59	60	61	62	63	63
	90th	113	114	115	117	119	120	121	73	74	75	76	77	78	78
	95th	117	118	119	121	123	124	125	77	78	79	80	81	82	82
	99th	124	125	127	129	130	132	132	85	86	87	88	89	90	90
12	50th	101	102	104	106	108	109	110	59	60	61	62	63	63	64
	90th	115	116	118	120	121	123	123	74	75	75	76	77	78	79
	95th	119	120	122	123	125	127	127	78	79	80	81	82	82	83
	99th	126	127	129	131	133	134	135	86	87	88	89	90	90	91

(cont.)

Table 1.2 (*Cont.*)

Age (year)	BP Percentile	Systolic BP (mm Hg) ←Percentile of Height→							Diastolic BP (mm Hg) ←Percentile of Height→						
		5th	10th	25th	50th	75th	90th	95th	5th	10th	25th	50th	75th	90th	95th
13	50th	104	105	106	108	110	111	112	60	60	61	62	63	64	64
	90th	117	118	120	122	124	125	126	75	75	76	77	78	79	79
	95th	121	122	124	126	128	129	130	79	79	80	81	82	83	83
	99th	128	130	131	133	135	136	137	87	87	88	89	90	91	91
14	50th	106	107	109	111	113	114	115	60	61	62	63	64	65	65
	90th	119	120	121	122	124	125	125	77	77	77	78	79	80	80
	95th	123	123	125	126	127	129	129	81	81	81	82	83	84	84
	99th	130	131	132	133	135	136	136	88	88	89	90	90	91	92
15	50th	107	108	109	110	111	113	113	64	64	64	65	66	67	67
	90th	120	121	122	123	125	126	127	78	78	78	79	80	81	81
	95th	124	125	126	127	129	130	131	82	82	82	83	84	85	85
	99th	131	132	133	134	136	137	138	89	89	90	91	91	92	93
16	50th	108	108	110	111	112	114	114	64	64	65	66	66	67	68
	90th	121	122	123	124	126	127	128	78	78	79	80	81	81	82
	95th	125	126	127	128	130	131	132	82	82	83	84	85	85	86
	99th	132	133	134	135	137	138	139	90	90	90	91	92	93	93
17	50th	108	109	110	111	113	114	115	64	65	65	66	67	67	68
	90th	122	122	123	125	126	127	128	78	79	79	80	81	81	82
	95th	125	126	127	129	130	131	132	82	83	83	84	85	85	86
	99th	133	133	134	136	137	138	139	90	90	91	92	93	93	93

The height percentiles are based on data available online at http://www.cdc.gov/growthcharts/. Adapted from National High Blood Pressure Education Program Working Group on High Blood Pressure in Children and Adolescents. The fourth report on the diagnosis, evaluation, and treatment of high blood pressure in children and adolescents. Pediatrics 2004; 114:555–576.

This is a work of the US government, published in the public domain by the American Academy of Pediatrics, available online at http://pediatrics.aappublications.org/cgi/content/full/114/2/S2/555 and http://www.nhlbi.nih.gov/health/prof/heart/hbp/hbp_ped.pdf.

Table 1.3 Blood Pressure Levels for Girls by Age (1–17 years) and Height Percentile.

Age (year)	BP Percentile	Systolic BP (mm Hg) ←Percentile of Height→							Diastolic BP (mm Hg) ←Percentile of Height→						
		5th	10th	25th	50th	75th	90th	95th	5th	10th	25th	50th	75th	90th	95th
1	50th	83	84	85	86	88	89	90	38	39	39	40	41	41	42
	90th	97	97	98	100	101	102	103	52	53	53	54	55	55	56
	95th	100	101	102	104	105	106	107	56	57	57	58	59	59	60
	99th	108	108	109	111	112	113	114	64	64	65	65	66	67	67
2	50th	85	85	87	88	89	91	91	43	44	44	45	46	46	47
	90th	98	99	100	101	103	104	105	57	58	58	59	60	61	61
	95th	102	103	104	105	107	108	109	61	62	62	63	64	65	65
	99th	109	110	111	112	114	115	116	69	69	70	70	71	72	72
3	50th	86	87	88	89	91	92	93	47	48	48	49	50	50	51
	90th	100	100	102	103	104	106	106	61	62	62	63	64	64	65
	95th	104	104	105	107	108	109	110	65	66	66	67	68	68	69
	99th	111	111	113	114	115	116	117	73	73	74	74	75	76	76
4	50th	88	88	90	91	92	94	94	50	50	51	52	52	53	54
	90th	101	102	103	104	106	107	108	64	64	65	66	67	67	68
	95th	105	106	107	108	110	111	112	68	68	69	70	71	71	72
	99th	112	113	114	115	117	118	119	76	76	76	77	78	79	79
5	50th	89	90	91	93	94	95	96	52	53	53	54	55	55	56
	90th	103	103	105	106	107	109	109	66	67	67	68	69	69	70
	95th	107	107	108	110	111	112	113	70	71	71	72	73	73	74
	99th	114	114	116	117	118	120	120	78	78	79	79	80	81	81
6	50th	91	92	93	94	96	97	98	54	54	55	56	56	57	58
	90th	104	105	106	108	109	110	111	68	68	69	70	70	71	72
	95th	108	109	110	111	113	114	115	72	72	73	74	74	75	76
	99th	115	116	117	119	120	121	122	80	80	80	81	82	83	83

(cont.)

Table 1.3 (Cont.)

Age (year)	BP Percentile	Systolic BP (mm Hg) ←Percentile of Height→							Diastolic BP (mm Hg) ←Percentile of Height→						
		5th	10th	25th	50th	75th	90th	95th	5th	10th	25th	50th	75th	90th	95th
7	50th	93	93	95	96	97	99	99	55	56	56	57	58	58	59
	90th	106	107	108	109	111	112	113	69	70	70	71	72	72	73
	95th	110	111	112	113	115	116	116	73	74	74	75	76	76	77
	99th	117	118	119	120	122	123	124	81	81	82	82	83	84	84
8	50th	95	95	96	98	99	100	101	57	57	57	58	59	60	60
	90th	108	109	110	111	113	114	114	71	71	71	72	73	74	74
	95th	112	112	114	115	116	118	118	75	75	75	76	77	78	78
	99th	119	120	121	122	123	125	125	82	82	83	83	84	85	86
9	50th	96	97	98	100	101	102	103	58	58	58	59	60	61	61
	90th	110	110	112	113	114	116	116	72	72	72	73	74	75	75
	95th	114	114	115	117	118	119	120	76	76	76	77	78	79	79
	99th	121	121	123	124	125	127	127	83	83	84	84	85	86	87
10	50th	98	99	100	102	103	104	105	59	59	59	60	61	62	62
	90th	112	112	114	115	116	118	118	73	73	73	74	75	76	76
	95th	116	116	117	119	120	121	122	77	77	77	78	79	80	80
	99th	123	123	125	126	127	129	129	84	84	85	86	86	87	88
11	50th	100	101	102	103	105	106	107	60	60	60	61	62	63	63
	90th	114	114	116	117	118	119	120	74	74	74	75	76	77	77
	95th	118	118	119	121	122	123	124	78	78	78	79	80	81	81
	99th	125	125	126	128	129	130	131	85	85	86	87	87	88	89

Age	%ile	SBP							DBP						
12	50th	102	103	104	105	107	108	109	61	61	61	62	63	64	64
	90th	116	116	117	119	120	121	122	75	75	75	76	77	78	78
	95th	119	120	121	123	124	125	126	79	79	79	80	81	82	82
	99th	127	127	128	130	131	132	133	86	86	86	88	88	89	90
13	50th	104	105	106	107	109	110	110	62	62	62	63	64	65	65
	90th	117	118	119	121	122	123	124	76	76	76	77	78	79	79
	95th	121	122	123	124	126	127	128	80	80	80	81	82	83	83
	99th	128	129	130	132	133	134	135	87	87	87	88	89	90	91
14	50th	106	106	107	109	110	111	112	63	63	63	64	65	66	66
	90th	119	120	121	122	124	125	125	77	77	77	78	79	80	80
	95th	123	123	125	126	127	129	129	81	81	81	82	83	84	84
	99th	130	131	132	133	135	136	136	88	88	88	89	90	91	92
15	50th	107	108	109	110	111	113	113	64	64	64	65	66	67	67
	90th	120	121	122	123	125	126	127	78	78	78	79	80	81	81
	95th	124	125	126	127	129	130	131	82	82	82	83	84	85	85
	99th	131	132	133	134	136	137	138	89	89	89	90	91	92	93
16	50th	108	108	110	111	112	114	114	64	64	65	66	66	67	68
	90th	121	122	123	124	126	127	128	78	78	79	80	81	81	82
	95th	125	126	127	128	130	131	132	82	82	83	84	85	85	86
	99th	132	133	134	135	137	138	139	90	90	90	91	92	93	93
17	50th	108	109	110	111	113	114	115	64	65	66	66	67	67	68
	90th	122	122	123	125	126	127	128	78	79	80	80	81	81	82
	95th	125	126	127	129	130	131	132	82	83	84	84	85	85	86
	99th	133	133	134	136	137	138	139	90	91	92	91	92	93	93

appropriate-sized cuff, usually larger than the cuff used for measurement of the arm blood pressure in the same patient. Since the same-sized cuff is frequently used at both sites, the pressure values obtained may be higher in the legs than in the arms. Coarctation of the aorta is suspected when the systolic pressure is 20 mm Hg lower in the legs than in the arms.

Blood pressure must be recorded properly by listing on the patient's record the systolic and diastolic pressure values, the method of obtaining the pressure, the extremity used, and whether upper- and lower-extremity blood pressures were done simultaneously or sequentially. When using automated methods requiring nonsimultaneous measurement, recording the heart rate measured with each pressure reading may be helpful, since wide rate variations may give a clue to varying states of anxiety and may help in the interpretation of differing pressure values.

Pulse pressure. Pulse pressure (the difference of the systolic and diastolic pressures) normally should be approximately one third of the systolic pressure. A narrow pulse pressure is associated with a low cardiac output or severe aortic stenosis. Pulse pressure widens in conditions with an elevated cardiac output or with abnormal runoff of blood from the aorta during diastole. The former occurs in such conditions as anemia and anxiety, whereas the latter is found in patients with conditions such as patent ductus arteriosus or aortic insufficiency.

Pulse
In palpating the child's pulse, not only the rate and rhythm but also the quality of the pulse should be carefully noted, as the latter reflects pulse pressure. Brisk pulses reflect a widened pulse pressure, whereas weak pulses indicate reduced cardiac output and/or narrowed pulse pressure. Coarctation of the aorta, for example, can be considered from a comparison of the femoral with the upper-extremity pulses. Mistakes have been made, however, in interpreting the quality of femoral arterial pulses. Palpation alone is not sufficient either to diagnose or to rule out coarctation of the aorta. Blood pressures must be taken in both arms and one leg.

Respiratory rate and effort
The respiratory rate and respiratory effort should be noted. Normal values for the respiratory rate are given in Table 1.4. Although the upper limit of normal respiratory rate for an infant is frequently given as 40 breaths per minute, observed rates can be as high as 60 breaths per minute in normal infants; the respiratory effort in such infants is easy. Difficulty with breathing is indicated by intercostal or suprasternal retractions or by flaring of the alae nasae. Premature infants or neonates may show periodic breathing, so the rate should be counted for a full minute.

Table 1.4 Normal Respiratory Rates at Different Ages.*

Age	Rates
Birth	30–60 (35)
First year	30–60 (30)
Second year	25–50 (25)
Adolescence	15–30 (15)

*Respiratory rates (breaths/min) vary with changes in mental state and physical activity. Sleeping rates are slower and are indicated in parentheses. Depth of respirations and effort expended by the patient are equally or more important than the rate itself.

Cardiac examination
Inspection
Cardiac examination begins with inspection of the thorax. A precordial bulge may be found along the left sternal border in children with cardiomegaly. The upper sternum may bulge in children with a large left-to-right shunt and pulmonary hypertension or with elevated pulmonary venous pressure.

Palpation
Several findings may be discovered by palpation; the most important is the location of the cardiac apex, an indicator of cardiac size. Obviously, if the apex is in the right hemithorax, there is dextrocardia.

Point of maximal impulse. In infants and children under 4 years of age, the apex impulse, which is the most lateral place that the cardiac impulse can be palpated, should be located in the fourth intercostal space at the midclavicular line. In older children it is located in the fifth intercostal space at the midclavicular line. Displacement laterally or inferiorly indicates cardiac enlargement.

Thrills. These are best identified by palpation of the precordium with the palmar surfaces of the metacarpophalangeal and proximal interphalangeal joints. Thrills are coarse, low-frequency vibrations occurring with a loud murmur, and are located in the same area as the maximal intensity of the murmur. In any patient suspected of congenital heart disease, the suprasternal notch also should be palpated but with a fingertip. A thrill at this site indicates a murmur originating from the base of the heart, most commonly aortic stenosis, less commonly pulmonary stenosis. In patients with patent ductus arteriosus or aortic insufficiency, the suprasternal notch is very pulsatile.

Heaves. Forceful, outward movements of the precordium (heaves) indicate ventricular hypertrophy. Right ventricular heaves are located along the right sternal border, and left ventricular heaves are located at the cardiac apex.

Percussion

Percussion of the heart can substantiate estimation of cardiac size in addition to that obtained by inspection and palpation.

Auscultation of the heart

Auscultation of the heart provides perhaps the most useful diagnostic information and should be performed in a systematic way so as to obtain optimum information.

Instrumentation. A good stethoscope is a must. It should have short, thick tubing; snug-fitting earpieces; and both a bell and a diaphragm. Low-pitched sounds and murmurs are heard best with the bell, and high-pitched sounds with the diaphragm. For most children a 3/4-inch bell and a 1-inch diaphragm are suitable for auscultation, although an adult-sized bell and diaphragm are preferable if adequate contact can be made with the chest wall. A 1-inch in diameter, diaphragm can be used in all aged children, since only part of the diaphragm need be in contact with the chest wall to transmit sound. Smaller sized diaphragms provide poor sound transmission.

Position and technique. In infants, initially auscultate through the clothing despite the often-quoted admonition that auscultation should never be performed in such a manner. Sometimes removing the clothes disturbs the child and results in a fussy state that precludes adequate auscultation. After the initial measurement, the clothing can be removed for another listen. Make certain the chest pieces of the stethoscope are warm.

With children between the ages of 1 and 3 years, listening is easier if they are sitting in their parent's lap because children of this age are usually frightened by strangers. In older children, the physical examination can proceed as in adults.

When auscultating, sitting alongside the child is easiest. This position is neither fatiguing to the examiner nor threatening to the child.

Auscultation of the heart should proceed in an orderly, stepwise fashion. Both the anterior and posterior thorax are auscultated with the patient in the upright position. Then the precordium is reexamined with the patient reclining. Each of the five major areas (aorta, pulmonary, tricuspid, mitral, and back) is carefully explored. Both the bell and diaphragm should be used in auscultation of the heart. High-pitched murmurs and the first and second heart sounds are heard better with the diaphragm; low-pitched murmurs and the third heart sound are most evident with the bell. The diaphragm should be applied with

moderate pressure; the bell must be applied with only enough pressure for uniform contact but not enough force to stretch the underlying skin into a "diaphragm," which alters the sensitivity to low frequencies. When auscultating the heart, attention is directed not only to cardiac murmurs but also to the quality and characteristics of the heart sounds.

Physiologic basis of auscultation. The events and phases of the cardiac cycle should be reviewed. Figure 1.2 represents a modification of a diagram by Wiggers and shows the relationship between cardiac pressures, heart sounds, and electrocardiogram. In studying this diagram, relate the events vertically as well as horizontally.

Systole. The onset of ventricular systole occurs following depolarization of ventricles and is indicated by the QRS complex of the electrocardiogram. As the ventricles begin to contract, the papillary muscles close the mitral and tricuspid valves. The pressure in the ventricles soon exceeds the atrial pressure and continues to rise until it reaches the diastolic pressure in the great vessel, at which point the semilunar valves open. The period of time between closure of the atrioventricular (AV) valves and the opening of the semilunar valves represents the *isovolumetric contraction period.* During this period, blood neither enters nor leaves the ventricles. During the next period, the ejection period, blood leaves the ventricles, and the ventricular pressure slightly exceeds the pressure in the corresponding great artery. As blood flow decreases, eventually the pressure in the ventricle falls below that in the great vessel, and the semilunar valve closes. This point represents the end of systole. The pressure in the ventricles continues to fall until it reaches the pressure of the corresponding atrium, at which time the AV valve opens. The period between closure of the semilunar valves and the opening of the AV valves is termed the *isovolumetric relaxation period* because blood neither enters nor leaves the ventricles.

Diastole. Diastole is divided into three consecutive phases:
Early. Early diastole is defined as the portion of ventricular diastole comprising the isovolumetric relaxation period, a time when ventricular pressures are falling but the volume is not changing because all cardiac valves are closed.

Mid. Mid-diastole begins with the opening of the AV valves; 80% of the cardiac output traverses the AV valves during mid-diastole. It has two distinct phases, a rapid and a slow filling phase. The rapid filling phase

22   Pediatric cardiology
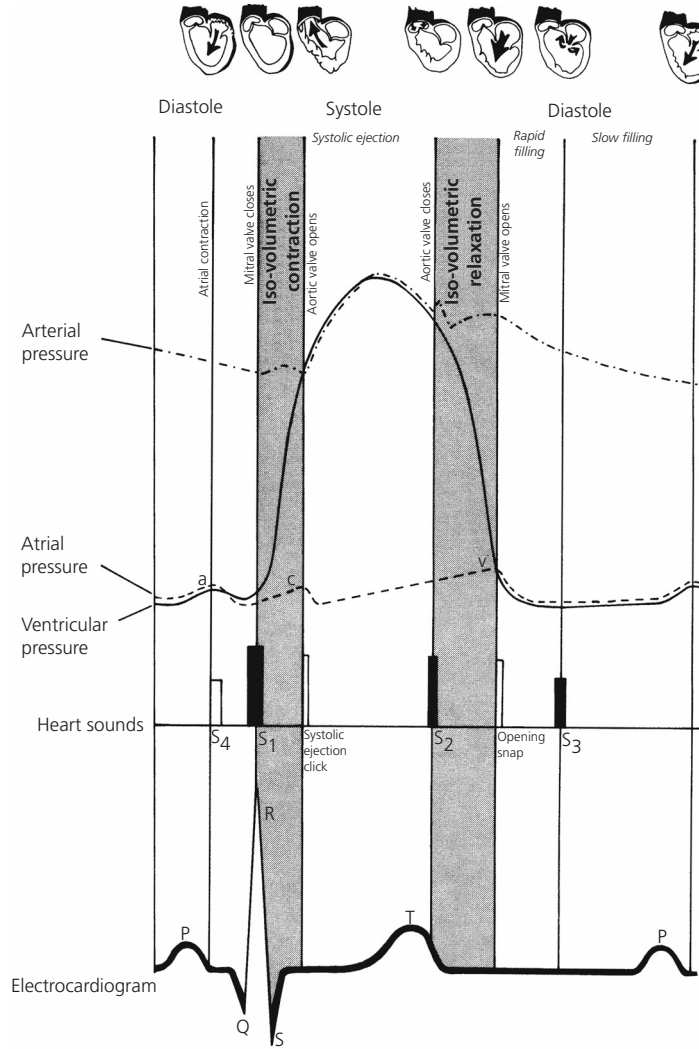
Figure 1.2 Relationship between cardiac pressures, electrocardiogram, heart sounds, and phases of the cardiac cycle. (Abbreviations: S_1, first heart sound; S_2, second heart sound, etc.)

comprises approximately the first 20% of diastole, during which about 60% of blood flow into the ventricle occurs. When a third heart sound (S_3) is present, it occurs at the boundary between the rapid and slow filling phases (see Fig. 1.2).

Late. Late-diastole begins with atrial contraction and the remaining 20% of ventricular filling occurs.

Interpretation of cardiac sounds and murmurs. The timing and meaning of cardiac sounds and murmurs are easily understood by considering their location within the cardiac cycle and the corresponding cardiac events. Although the origin of certain heart sounds remains controversial, in this chapter we discuss them as originating from valvar events.

Heart sounds. The first heart sound (S_1) represents closure of the mitral and tricuspid valves (Fig. 1.2) and occurs as the ventricular pressure exceeds the atrial pressure at the onset of systole. In children, the individual mitral and tricuspid components are usually indistinguishable, so the first heart sound appears single. Occasionally, two components of this sound are heard. Splitting of the first heart sound can be a normal finding, although patients with complete right bundle branch block show wide splitting, since tricuspid valve closure is delayed secondary to delayed right ventricular activation.

The first heart sound is soft if prolonged AV conduction occurs, which allows the valves to drift closed after atrial contraction, or if myocardial disease is present.

The first heart sound is accentuated in conditions with increased blood flow across an AV valve (as in left-to-right shunt) or in high cardiac output.

The second heart sound (S_2) is of great diagnostic significance, particularly in a child with a cardiac malformation. The normal second heart sound has two components which represent the asynchronous closure of the aortic and pulmonary valves. These sounds signal the completion of ventricular ejection. Aortic valve closure normally precedes closure of the pulmonary valve because right ventricular ejection is longer. The presence of the two components, aortic (A_2) and pulmonic (P_2), is called splitting of the second heart sound (Fig. 1.3).

The time interval between the components varies with respiration. Normally, on inspiration the degree of splitting increases, whereas on expiration it shortens. This variation is related to the greater volume of blood that returns to the right side of the heart during inspiration. Since ejection of this augmented volume of blood requires a longer time, the second heart sound becomes more widely split on inspiration.
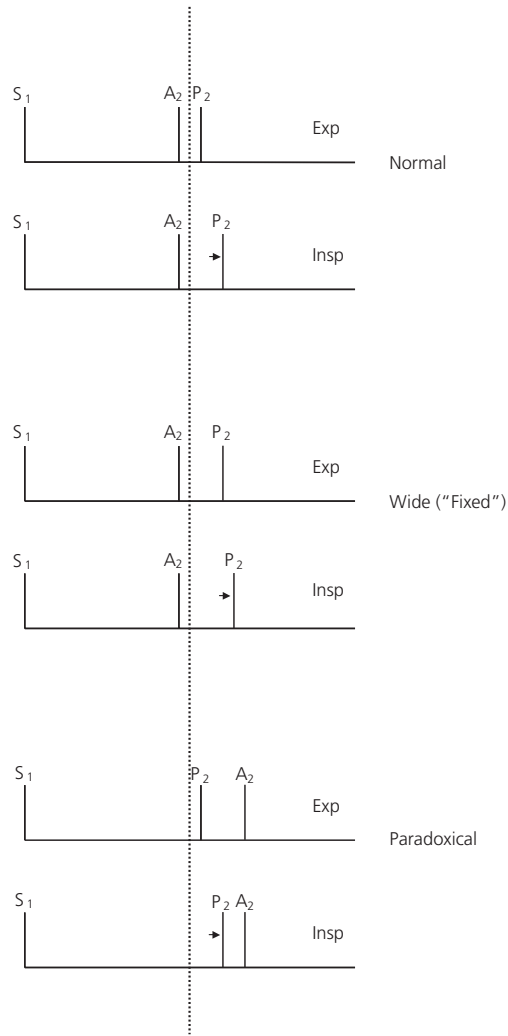
Figure 1.3 Respiratory variations in splitting of second heart sound. In a normal individual, P_2 (pulmonary component of second heart sound) is delayed on inspiration. Wide splitting occurs in conditions prolonging right ventricular ejection. Paradoxical splitting occurs in conditions delaying A_2 (aortic component of second heart sound). P_2 changes normally with inspiration. Thus, the interval between P_2 and A_2 narrows on inspiration and widens on expiration.

> The second heart sound can be split abnormally:
> *Wide splitting.* Conditions prolonging right ventricular ejection lead to wide splitting of the second heart sound because P_2 is delayed further than normal. This phenomenon is present in three hemodynamic states.
>   Conditions in which the right ventricle ejects an increased volume of blood (e.g., ASD–but not VSD).
> Obstruction to right ventricular outflow (e.g., pulmonary stenosis).
>   Delayed depolarization of the right ventricle (e.g., complete right bundle branch block).
>
> *Paradoxical splitting.* Paradoxical splitting of the second heart sound is probably of greater importance in understanding the physiology of heart sounds than in reaching a cardiac diagnosis in children. Conditions prolonging left ventricular ejection may delay the aortic component causing it to follow the pulmonary component (Fig. 1.3). Thus, as P_2 varies normally with respiration, the degree of splitting widens paradoxically on expiration and narrows on inspiration. Left ventricular ejection is prolonged in conditions in which the left ventricle ejects an increased volume of blood into the aorta (e.g., patent ductus arteriosus), in left ventricular outflow obstruction (e.g., aortic stenosis), and in delayed depolarization of the left ventricle (complete left bundle branch block).

Thus, wide splitting and paradoxical splitting of the second heart sound occur from similar cardiac abnormalities but on opposite sides of the heart. Paradoxical splitting is associated with severe left-sided disorders.

Intensity of P_2. In assessing a child with a cardiac anomaly, particular attention also should be directed toward the intensity of the pulmonary component (P_2) of the second heart sound. The pulmonic component of the second sound is accentuated whenever the pulmonary arterial pressure is elevated, whether this elevation is related to pulmonary vascular disease or to increased pulmonary arterial blood flow. In general, as the level of pulmonary arterial pressure increases, the pulmonic component of the second heart sound becomes louder and closer to the aortic component.

Single second heart sound. The finding of a single second heart sound usually indicates that one of the semilunar valves is atretic or severely stenotic because the involved valve does not contribute its component to the second sound. The second heart sound also is single in patients with persistent truncus arteriosus because there is only a single semilunar valve or whenever pulmonary arterial pressure is at systemic levels, and the aortic and pulmonary artery pressure curves are superimposed.

Third heart sound (S_3) may be present in a child without a cardiac anomaly but may be accentuated in pathologic states. This sound occurs early in diastole

and represents the transition from rapid to slow filling phases. In conditions with increased blood flow across either the mitral valve (as in mitral insufficiency) or the tricuspid valve (as in ASD), the third heart sound may be accentuated. A gallop rhythm found in congestive cardiac failure often represents exaggeration of the third heart sound in the presence of tachycardia.

Fourth heart sounds (S_4) are abnormal. Located in the cardiac cycle late in diastole, they occur with the P wave of the electrocardiogram and exist synchronous to the atrial "a" wave. They are found in conditions in which either the atrium forcefully contracts against a ventricle with decreased compliance, as from fibrosis or marked hypertrophy, or when the flow from the atrium to the ventricle is greatly increased. The fourth heart sound may be audible as a presystolic gallop, particularly if tachycardia is present.

Systolic ejection clicks are abnormal and occur at the time the semilunar valves open. Therefore, they mark the transition from the isovolumetric contraction period to the onset of ventricular ejection. Ordinarily this event is not heard, but in specific cardiac conditions, a sound (systolic ejection click) may be present at this point in the cardiac cycle and because of its timing be confused with a split first heart sound.

Systolic ejection clicks indicate the presence of a dilated great vessel, most frequently from poststenotic dilation. These sharp, high-pitched sounds have a clicky quality. Ejection clicks of aortic origin are heard best at the cardiac apex or over the left lower thorax when the patient is in a supine position; they vary little with respiration. Aortic ejection clicks are common in patients with valvar aortic stenosis or a bicuspid aortic valve with concomitant poststenotic dilation. Ejection clicks may also originate from a dilated pulmonary artery, as present in pulmonary valvar stenosis or pulmonary arterial hypertension. Pulmonic ejection clicks are best heard in the pulmonary area when the patient is sitting; they vary in intensity with respiration. Ejection clicks in patients with a stenotic semilunar valve occur more commonly in mild or moderate cases; they may be absent in patients with severe stenosis.

Clicks are not associated with subvalvar stenosis since there is no poststenotic dilation.

Opening snaps are abnormal and occur when AV valves open. At this point, the ventricular pressure is falling below the atrial pressure, the isovolumetric relaxation period is ending, and ventricular filling is beginning. Ordinarily, no sound is heard at this time, but if the AV valve is thickened or fibrotic, a low-pitched noise may be heard when it opens. Opening snaps, rare in children, are almost always associated with rheumatic mitral valvar stenosis.

Murmurs. Cardiac murmurs are generated by turbulence in the normal laminar blood flow through the heart. Turbulence results from narrowing the pathway of blood flow, abnormal communications, or increased blood flow.
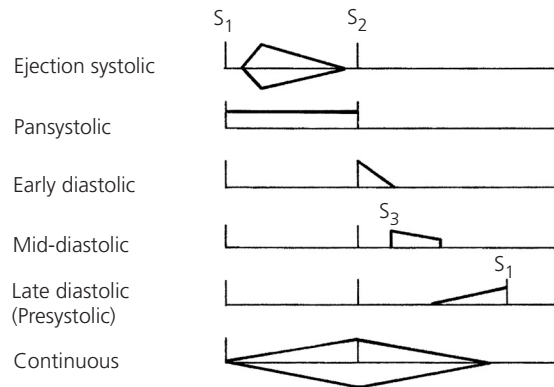
Figure 1.4 Classification of murmurs, showing location within cardiac cycle and usual contour. (Abbreviations: S_1, first heart sound; S_2, second heart sound; S_3, third heart sound.)

Five aspects of a cardiac murmur provide knowledge of the underlying cause of turbulence: location in cardiac cycle (timing), location on thorax, radiation of murmur, loudness, and pitch and character.

Location in cardiac cycle (timing). Murmurs may be classified by their location within the cardiac cycle (Fig. 1.4). A murmur is heard only during that portion of the cardiac cycle in which turbulent blood flow occurs.

Systolic murmurs. Two types of systolic murmurs exist: holosystolic and systolic ejection.

Holosystolic murmurs (synonyms are pansystolic or systolic regurgitant) start with the first heart sound and continue into systole, often extending to the second heart sound. Therefore, these murmurs involve the isovolumetric contraction period.

Only two conditions permit blood flow during isovolumetric contraction:
   Ventricular septal defect.
   Atrioventricular valve insufficiency (mitral, tricuspid, or the "common" valve in AV canal defect).

In VSD, flow occurs between the left and right ventricles from the onset of systole, whereas in AV valve insufficiency the high-pressure ventricle is in communication with the lower-pressure atrium from the time of the first heart sound.

Because holosystolic murmurs begin so close to the first heart sound, that sound may be masked at the location of maximal murmur intensity. This masking can be a clue to a holosystolic murmur, particularly in patients with rapid heart rate.

Systolic ejection murmur (SEM) results from turbulent forward blood flow across a semilunar valve (aortic, pulmonary, or truncal valve), a great vessel, or ventricular outflow tract. Since turbulent flow in these locations cannot begin until the semilunar valves open, an interval (the isovolumetric contraction period) exists between the first heart sound and the onset of the murmur. Although often diamond-shaped (crescendo/decrescendo), SEMs are distinguished by the delayed onset of the murmur after the isovolumetric contraction period.

Ejection murmurs are found in such conditions as ASD, aortic stenosis, and pulmonary stenosis. In contrast to holosystolic murmurs, the first heart sound is distinctly audible at the site where the SEM is best heard.

Diastolic murmurs can also be classified according to their timing in the cardiac cycle.

Early diastolic murmurs occur immediately following the second heart sound and include the isovolumetric relaxation period. During this time, blood can only flow from a higher-pressure great vessel into a lower-pressure ventricle.

> Early diastolic murmurs indicate regurgitation across a semilunar valve (aortic, pulmonary, or truncal valve insufficiency).

Usually decrescendo, their pitch depends on the level of diastolic pressure within the great vessel: high-pitched in aortic or truncal regurgitation and lower-pitched with pulmonary regurgitation (unless pulmonary hypertension is present).

Mid-diastolic murmurs (sometimes called inflow murmurs) occur at the time of maximal passive ventricular filling and usually result from increased forward blood flow across a normal AV valve. In children, they occur most commonly in conditions with increased pulmonary blood flow and therefore, with increased blood flow into the ventricles (as in ASD or VSD). These low-pitched rumbles are usually heard only with the bell of the stethoscope and are easily overlooked by an inexperienced examiner.

Late diastolic murmurs represent organic obstruction of an AV valve. These murmurs crescendo with a low pitch. Rheumatic mitral stenosis is the typical example.

Table 1.5 Characteristics of Murmurs.

Location in Cardiac Cycle	Type of murmur	
	Regurgitant	Forward Flow

Systolic	Holosystolic
	  Begins with S_1
	  Includes isovolumetric
	    contraction period
		Ejection
		  Follows S_1
		  Occurs after isovolumetric
		    contraction period

Diastolic	Early diastolic
	  Begins with S_2
	  Includes isovolumetric
	    relaxation period
		Mid- or late diastolic
		  Follows S_2
		  Occurs after isovolumetric
		    relaxation period

Continuous	Systole and diastole Continues during S_2	

S_1, first heart sound; S_2, second heart sound.

Continuous murmur. A continuous murmur indicates turbulence beginning in systole and extending into diastole. It may last throughout the cardiac cycle. Usually, it occurs when communication exists between the aorta and the pulmonary artery or other portions of the venous side of the heart or circulation.

Patent ductus arteriosus is the classic example, but continuous murmurs are heard with other types of systemic arteriovenous fistulae.

The similarities and differences between regurgitant murmurs and those due to forward blood flow, whether in systole or diastole, are summarized in Table 1.5.

Regurgitant murmurs begin with either the first or second heart sound and include the isovolumetric periods, whereas those related to abnormalities of forward flow begin after an isovolumetric period and may be associated with an abnormal cardiac sound (systolic ejection click or opening snap). A notable exception to these rules is the murmur associated with mitral valve prolapse, discussed in Chapter 10. Table 1.6 presents differential diagnosis of murmurs by timing.

Location on the thorax. The location of the maximal intensity of murmurs on the thorax (Fig. 1.5) provides information about the anatomic origin of the murmur:

(a) *Aortic area*: from the mid-left sternal border to beneath the right clavicle.
(b) *Pulmonary area*: the upper left sternal border and beneath the left clavicle.
(c) *Tricuspid area*: along the lower left and right sternal border.
(d) *Mitral area*: the cardiac apex.

30 Pediatric cardiology

Table 1.6 Differential Diagnosis of Murmurs by Location in Cardiac Cycle.

Location in Cardiac Cycle	Timing	Physiology	Possible Conditions
Systolic	Holosystolic	Flow, ventricle to ventricle Regurgitation, ventricle to atrium	VSD AV valve regurgitation (MR, TR, common AV valve regurgitation)
	Ejection	Flow, ventricle to artery	Semilunar valve, outflow tract, or branch pulmonary artery flow (normal) Increased pulmonary valve flow (e.g. ASD, AVM—abnormal)
		Stenosis, ventricle to artery	Semilunar valve stenosis (e.g., AS, PS, truncal valve stenosis), subvalvar stenosis, or supravalvar stenosis
	Mid- to late systolic	Regurgitation, ventricle to atrium, only with AV valve prolapse	Mitral valve prolapse with regurgitation
Diastolic	Early diastolic	Regurgitation, artery to ventricle	Semilunar valve regurgitation (AI, PI, truncal valve insufficiency)
	Mid- or late diastolic	Flow, atrium to ventricle	Increased flow via AV valve (e.g. Mitral mid-diastolic murmur in VSD, PDA, or severe MR; Tricuspid valve mid-diastolic murmur in ASD, AVM)
		Stenosis, atrium to ventricle	AV valve stenosis (e.g., MS, TS)
Continuous	Systolic accentuation	Flow, artery to artery	PDA Surgical systemic artery to pulmonary artery shunt
		Flow, artery to vein	AVM
		Flow, within artery	Arterial bruit
	Respiratory accentuation	Flow, within vein	Venous hum

AI, aortic insufficiency (regurgitation); AS, aortic stenosis; ASD, atrial septal defect; AV, atrioventricular; AVM, arteriovenous malformation; MR, mitral regurgitation; MS, mitral stenosis; PDA, patent ductus arteriosus; PI, pulmonary insufficiency (or regurgitation); PS, pulmonary stenosis; TR, tricuspid regurgitation; TS, tricuspid stenosis; VSD, ventricular septal defect.
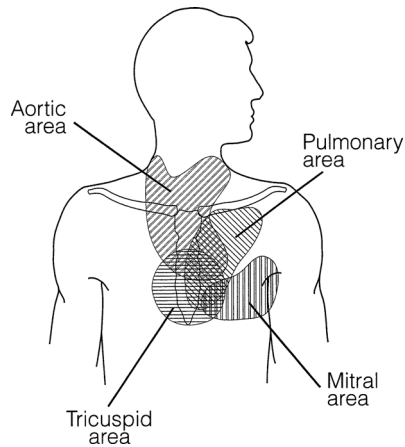
Figure 1.5 Primary areas of auscultation. (Reprinted from Pediatr Clin North Am, Vol 45, pelech AN, The cardiac murmur: When to refer? Pages 107–122, Copyright 1998, with permission from Elsevier.)

In these areas respectively, the murmurs of aortic stenosis, pulmonary stenosis, tricuspid insufficiency, and mitral insufficiency are found. In infants and children, listening over both sides of the back is essential. For example, the murmur of coarctation of the aorta is heard best in the left paraspinal area, directly over the anatomic site of the aortic narrowing. The murmur of peripheral pulmonary artery stenosis is heard over both sides of the back and axillae.

Radiation of murmurs. The direction of transmission of the murmur is also helpful, for it reflects the direction of turbulent flow, which often is along major blood vessels.

Murmurs originating from the aortic outflow area (e.g., aortic valvar stenosis) radiate toward the neck and into the carotid arteries.

Murmurs from the pulmonary outflow area are transmitted to the left upper back.

Mitral murmurs are transmitted toward the cardiac apex and left axilla; occasionally, mitral regurgitation is heard in the middle back.

Loudness. The loudness of a cardiac murmur is graded on a scale in which grade 6 represents the loudest murmur. Conventionally, loudness is indicated by a fraction in which the numerator indicates the loudness of the patient's murmur and the denominator indicates the maximum grade possible. Although somewhat arbitrary, the classification is based on sound intensity and chest wall vibration (thrills).

1/6 is very soft—heard only with careful attention
2/6 is not loud but is easily heard
3/6 is loud but no thrill can be palpated
4/6 is loud and associated with a thrill
5/6 is very loud
6/6 very loud—heard even with the stethoscope held just off the chest
    wall

The pitch of the murmur can be described as high, medium, or low. High-pitched murmurs (heard with a diaphragm) occur when a high-pressure difference in the turbulent flow exists, such as in aortic or mitral insufficiency. Low-pitched murmurs (heard with a bell) occur when there is a low-pressure difference, as in the mid-diastolic mitral inflow murmur accompanying a VSD.

The character of the murmur can be helpful in distinguishing certain causes. Harsh murmurs are typical of severe outflow stenosis when a large pressure difference is present, as in aortic valvar stenosis.

Normal murmurs. Distinction between a normal or functional (innocent) and a significant (organic) murmur can be difficult in some children. Although this text describes the characteristics of the commonly heard functional murmurs, only by experience and careful auscultation one can become proficient in distinguishing a functional murmur from a significant one.

Functional murmurs have four features which help to distinguish them from significant murmurs: (a) normal heart sounds, (b) normal heart size, (c) lack of significant cardiac signs and symptoms, and (d) loudness of grade 3/6 or less.

Some mild forms of cardiac abnormalities may have these features. Thus, the ability to categorize the murmur as a specific type of functional murmur is helpful.

Six types of normal or functional murmurs follow:
(1) *Still's murmur*. Often called "musical" or "twangy string," this soft (grade 1/6–3/6), low-pitched vibratory SEM is heard between the lower left sternal border and apex. Because of this location on the thorax, it may be misinterpreted as a VSD. It can be distinguished because it begins

after, not with, the first heart sound (as in VSD), and lacks the harsh quality of a VSD murmur.

(2) *Pulmonary flow murmur*. This soft (grade 1/6–3/6) low-pitched SEM is heard in the pulmonary area. The murmur itself may be indistinguishable from ASD. With this functional murmur however, the characteristics of the second heart sound remain normal, whereas in ASD the components of the second heart sound show wide, fixed splitting.

(3) *Normal neonatal pulmonary artery branch flow murmur*. This soft SEM is heard in many premature neonates, often at the time their physiologic anemia reaches its nadir, and in many term infants. It is characterized by a soft systolic flow murmur best heard in the axillae and back, and poorly heard, if at all, over the precordium. To avoid confusion with true pulmonary artery pathology, the synonym peripheral pulmonic stenosis, or PPS, should not be used.

(4) *Venous hum*. This murmur might be confused with a patent ductus arteriosus because it is continuous. It is heard best, however, in the right infraclavicular area. Venous hum originates from turbulent flow in the jugular venous system. Several characteristics distinguish it from patent ductus arteriosus: it can be louder in diastole and varies with respiration; it is best heard with the patient sitting; it diminishes and usually disappears when the patient reclines; and it changes in intensity with movements of the head or with pressure over the jugular vein.

(5) *Cervical bruit*. In children, a soft systolic arterial bruit may be heard over the carotid arteries. They are believed to originate at the bifurcation of the carotid arteries. The bruit should not be confused with the transmission of cardiac murmurs to the neck, as in aortic stenosis. Aortic stenosis is associated with a suprasternal notch thrill.

(6) *Cardiopulmonary murmur*. This sound (more along the mid left sternal border than right) originates from compression of the lung between the heart and the anterior chest wall. This murmur or sound occurs during systole, becomes louder in mid-inspiration and mid-expiration, and sounds close to the ear.

In most children with a functional cardiac murmur, a chest X-ray, electrocardiogram, or echocardiogram is unnecessary, as the diagnosis can be made with certainty from the physical examination alone. In a few patients these studies may be indicated to distinguish a significant and functional murmur. If it is a normal (innocent) murmur, the parents and the patient should be reassured of its benign nature. No special care is indicated for these children, and the child can be monitored at intervals dictated by routine pediatric care by their own medical provider. Many (not all) functional murmurs disappear in adolescence,

and the murmurs may be accentuated during times of increased cardiac output, such as during fever and anemia.

Abdominal examination. The abdomen should also be carefully examined for the location and size of the liver and spleen. The examiner should be alert to the presence of situs inversus. The hepatic edge should be palpated and its distance below the costal margin measured. If the edge is lower than normal, the upper margin of the liver should be percussed to determine the span of the liver. In patients with a depressed diaphragm (e.g., from asthma), the liver edge is also depressed downward; in this instance, the upper extent of the liver is also depressed. The liver edge normally is palpable until 4 years of age. Pulsatile motion may be palpated over the liver in severe tricuspid regurgitation or transmitted through soft tissues from a hyperdynamic heart in the absence of AV valve regurgitation.

The spleen ordinarily should not be palpable. It may be enlarged in patients with chronic congestive cardiac failure or infective endocarditis.

LABORATORY EXAMINATION

Electrocardiography

Electrocardiography plays an integral part in evaluation of a child with cardiac disease. It is most useful in reaching a diagnosis when combined with other patient data. The electrocardiogram permits assessment of the severity of many cardiac conditions by reflecting the anatomic changes of cardiac chambers resulting from abnormal hemodynamics imposed by the cardiac anomaly.

For example, left ventricular hypertrophy develops in patients with aortic stenosis. The electrocardiogram reflects the anatomic change; and the extent of electrocardiographic change roughly parallels the degree of hypertrophy, yielding information about the severity of the obstruction. However, a pattern of left ventricular hypertrophy is not diagnostic of aortic stenosis because other conditions, such as systemic hypertension or coarctation of the aorta, also cause anatomic left ventricular hypertrophy and the associated electrocardiographic changes. Occasionally, electrocardiographic patterns are specific enough for diagnosis of a particular cardiac anomaly (e.g., anomalous left coronary artery, tricuspid atresia, or endocardial cushion defect).

The electrocardiogram is used to assess cardiac rhythm disturbances (see Chapter 10) and electrolyte abnormalities. Ambulatory electrocardiography (24-hour electrocardiogram or "Holter monitor") is used for surveillance of subclinical arrhythmias, to access the range and variability of heart rate, and to document the rhythm during symptoms. When symptoms suspected of originating from arrhythmia occur less frequently than daily, an event monitor allows recording of brief (1–2 minutes) electrocardiograms during symptoms for later transmission via telephone.
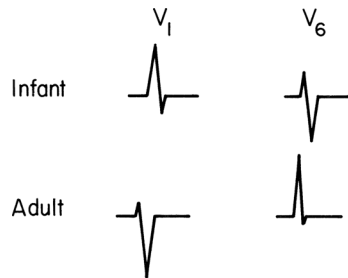
Figure 1.6 Comparison of the contour of QRS complex in leads V_1 and V_6 of infants and adults.

Developmental changes

The electrocardiogram of children normally changes with age; the greatest changes occur during the first year of life reflecting changes in the circulation. At birth the right ventricle weighs more than the left ventricle because during fetal life it supplied blood to the aorta by way of the ductus arteriosus and had a greater stroke volume than the left ventricle. As the child grows, the left ventricular wall thickens as systemic arterial pressure rises slowly; meanwhile, the right ventricular wall thins as pulmonary arterial pressure falls. These anatomic changes primarily affect those portions of the electrocardiogram reflecting ventricular depolarization (QRS complex) and repolarization (T waves).

Therefore, in infancy, the thicker than normal right ventricular wall directs the QRS axis more toward the right with tall R waves in lead V_1 and relatively deep S waves in lead V_6. With age, the QRS axis shifts toward the left, and leads V_1 and V_6 assume a pattern similar to that seen in adults (Fig. 1.6).

In interpreting the electrocardiogram of a child, these changes and others that occur with age must be considered. Table 1.7 shows the range of normal values for several electrocardiographic intervals and wave forms.

Technical factors

Analysis of an electrocardiogram should proceed in an orderly sequence to gain maximal information from the tracing. The speed and sensitivity of the recording should be noted, and variation from "standard" speed of 25 mm/s and amplitude of 10 mm/mV must be considered with comparison to normal values.

Rate and rhythm. The initial step should be to recognize any cardiac arrhythmias or major conduction abnormalities. These can usually be detected by answering the following three questions:

36 Pediatric cardiology

Table 1.7 Normal Values of Important Electrocardiographic Parameters.

Age	QRS Axis (degrees)	R Wave in V_1(mm)	S Wave in V_1(mm)	R Wave in V_6(mm)	S Wave in V_6(mm)
0–24 hours	137 (70–205)	16 (6–27)	10 (0–25)	4 (0–8)	4 (0–12)
1–7 days	125 (75–185)	17 (4–30)	10 (0–20)	6 (0–16)	3 (0–12)
8–30 days	108 (30–190)	13 (3–24)	7 (0–18)	8 (0–20)	2 (0–9)
1–3 months	75 (25–125)	10 (2–20)	7 (0–18)	9 (2–16)	2 (0–6)
3–6 months	65 (30–96)	10 (2–20)	7 (2–12)	10 (2–16)	1 (0–5)
6–12 months	65 (10–115)	10 (2–20)	8 (2–15)	12 (3–20)	1 (0–3)
1–3 years	55 (6–108)	9 (2–18)	10 (2–25)	12 (3–21)	1 (0–3)
3–5 years	62 (20–105)	7 (1–16)	13 (2–25)	13 (4–21)	1 (0–3)
5–8 years	65 (16–112)	7 (1–16)	14 (2–25)	14 (6–24)	1 (0–3)
8–12 years	62 (15–112)	6 (1–16)	14 (2–25)	14 (8–21)	1 (0–3)
12–16 years	65 (20–116)	5 (0–16)	15 (2–25)	13 (8–20)	1 (0–3)

Are there P waves?

Is each P wave followed by a QRS complex?

Is each QRS complex preceded by a P wave?

If the answer to any of these questions is no, the type of rhythm distur-bance should be further investigated by following the instructions given in Chapter 10.

Components of the electrocardiogram. The next step is the analysis of each component of the electrocardiographic tracing. This is accomplished not by looking at each lead from left to right, as in reading a newspaper but by reading up and down, first assessing the P waves in each lead, then at the QRS in each lead, and finally at the T wave in each lead.

For each wave form, four features are analyzed: axis, amplitude, duration, and any characteristic pattern (such as the delta wave of Wolf–Parkinson–White syndrome). Using groups of leads, axis is analyzed: the limb leads are used to derive frontal–plane axes, and the chest or precordial leads are used for horizontal plane axes.

Confusion exists about the word axis. Commonly the term "the axis" is used to describe the QRS in the standard leads. But, just as the direction of the QRS can be described so can that of P and T waves; the principle is the same.

P wave. The P wave is formed by depolarization of the atria. Depolarization is initiated from the sinoatrial node located at the junction of the superior vena cava and right atrium. It generally proceeds inferiorly and leftward toward the

AV node located at the junction of the atrium and ventricle, low in the right atrium and adjacent to the coronary sinus. The direction of atrial depolarization also proceeds slightly anteriorly. Since atrial depolarization begins in the right atrium, the initial portion of the P wave is formed primarily from right atrial depolarization, while the terminal portion is formed principally from left atrial depolarization.

The following three characteristics of the P wave should be studied.

(1) *P wave axis*. The P wave axis indicates the net direction of atrial depolarization (Fig. 1.7). Normally, the P wave axis in the frontal plane is $+60°$ ($+15°$ to $+75°$), reflecting the direction of atrial depolarization from the sinoatrial to the AV nodes.

> Therefore, the largest P waves are usually in lead II; the P waves are normally positive in leads I, II, and aVF; always negative in lead aVR; and positive, negative, or diphasic in leads aVL and III.
>
> In the horizontal plane, the P wave axis is directed toward the left (approximately lead V_5). Therefore, the P wave in lead V_1 may be positive, negative, or diphasic.

The P wave axis changes when the pacemaker initiating atrial depolarization is abnormally located. One example is mirror-image dextrocardia associated with situs inversus, in which the anatomic right atrium and the sinoatrial node are located on the left side, so atrial depolarization occurs from left to right. This leads to a P wave axis of $+120°$ with the largest P waves in lead III. Another example is junctional rhythm, in which atrial depolarization proceeds from the AV node in a superior-rightward direction.

(2) *P wave amplitude*. The P wave should not exceed 3 mm in height. Because most of the right atrium is depolarized before the left atrium, the early portion of the P wave is accentuated in right atrial enlargement.

> P waves taller than 3 mm indicate right atrial enlargement. This condition causes tall, peaked, and pointed P waves, usually found in the right precordial leads or in leads II, III, or aVF.

(3) *P wave duration*. The P wave should be less than 100 milliseconds in duration. When longer, left atrial enlargement or intraatrial block (much rarer) is present.

38 Pediatric cardiology

(a)

(b)

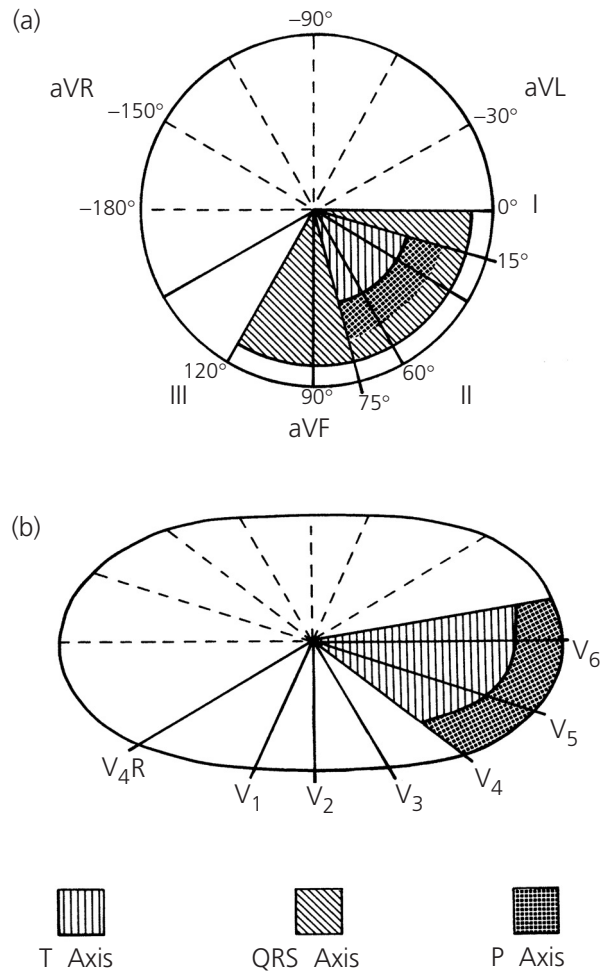Figure 1.7 is labeled with: T Axis, QRS Axis, P Axis

Figure 1.7 Electrocardiogram normal axes. Relationship of limb leads in frontal plane (a) and precordial leads in horizontal plane (b). The normal ranges for the P wave, QRS complex, and T wave axes in frontal plane and the P and T wave axes in horizontal plane are shown.

> In left atrial enlargement, the P wave is broad and notched,
> particularly in leads I, aVL, and/or leads V_5 and V_6; a broad negative
> component of the P wave may also exist in lead V_1 because the latter
> part of the P wave principally represents left atrial depolarization and
> because the left atrium faces the left precordial leads so the terminal
> P wave forces are accentuated and directed leftward.

PR interval. The PR interval is the time from the onset of the P wave to the
onset of the QRS complex. It represents the transmission of the impulse from
the sinoatrial node through the atria and then through the AV node and the
Purkinje system.

> The normal values of PR interval measured in leads I, II, or III are
> 100–120 milliseconds in infancy,
> 120–150 milliseconds in childhood, and
> 140–220 milliseconds in adulthood.
> However, the PR interval varies with heart rate as well as age, becoming
> shorter with faster rates.

A PR interval longer than these values is caused by prolongation of AV nodal
conduction, such as that caused by acute febrile illness or digoxin. The PR inter-
val may also be shorter than normal if an ectopic focus for atrial depolarization
exists, as in low atrial rhythm, or if an accessory conducting pathway into the
ventricle with preexcitation is present, as in Wolf–Parkinson–White syndrome.

QRS complex. The QRS complex represents ventricular depolarization. Ventric-
ular depolarization starts on the left side of the interventricular septum near
the base and proceeds across the septum from left to right. Depolarization of
the free walls of both ventricles follows. The posterior basilar part of the left
ventricle and the infundibulum of the right ventricle are the last portions of
ventricular myocardium to be depolarized.

   The QRS complex should be analyzed for the following features:

(1) *QRS axis*. The QRS axis represents the net direction of ventricular depolariza-
    tion. In children, the axis varies because of the hemodynamic and anatomic
    changes occurring with age. The value of the QRS axis in the frontal plane
    for various ages is shown in Table 1.7.

    In neonates, the QRS axis range is $+70°$ to $+215°$, but with age the
    axis comes into the range of $0°$ to $+120°$. Most of the change occurs by
    3 months of age (Fig. 1.7).

> Right axis deviation is diagnosed when the calculated value for the
> QRS axis is greater than the upper range of normal, which for older
> children is more than $+120°$. Right axis deviation is almost always
> associated with right ventricular hypertrophy or enlargement.
>
>   Left axis deviation is indicated when the calculated QRS axis is less
> than the smaller value of the normal range. Left axis deviation is
> associated with myocardial disease or ventricular conduction
> abnormalities, such as those that occur in endocardial cushion defect,
> but rarely with left ventricular hypertrophy.

When the QRS axis lies between $-90°$ and $-150°$ ($+210°$ to $+270°$),
deciding if this represents marked right axis deviation or marked left axis
deviation is difficult. In such patients, the practitioner should interpret the
location of the axis in light of the patient's cardiac anomaly.

Calculation of the direction of the mean QRS vector in the horizontal
plane is more difficult, but the vector can be generally described as ante-
rior, posterior, leftward, or rightward. Determination of the horizontal QRS
axis can be combined with information about QRS amplitude to determine
ventricular hypertrophy.

(2) *QRS amplitude*. In infants and children, little diagnostic information is ob-
tained from the QRS amplitude of the six standard leads except when low
voltage is present in these leads. Normally, the QRS complex in leads I, II,
and III exceeds 5 mm in height, but if smaller, suggests conditions such as
pericardial effusion.

In the precordial leads, QRS amplitude is used to determine ventricular hy-
pertrophy. Leads V_1 and V_6 should each exceed 8 mm; if smaller, pericardial
effusion or similar conditions may be present.

Ventricular hypertrophy is manifested by alterations in ventricular depo-
larization and amplitudes of the QRS complex. The term ventricular hyper-
trophy is partly a misnomer, as it applies to electrocardiographic patterns
in which the primary anatomic change is ventricular chamber enlargement
and to patterns associated with cardiac conditions in which the ventricular
walls are thicker than normal.

> Generally, hypertrophy is the response to pressure loads upon the
> ventricle (e.g., aortic stenosis), whereas enlargement reflects
> augmented ventricular volume (e.g., aortic insufficiency).

Interpretation of an electrocardiogram for ventricular hypertrophy must be
made relative to the normal evolution of the QRS complex, particularly to
the amplitude of the R and S waves in leads V_1 and V_6 (Table 1.7).

Right ventricular hypertrophy. In right ventricular hypertrophy, the major QRS forces are directed anteriorly and rightward. This usually leads to right axis deviation, a taller than normal R wave in lead V_1 and a deeper than normal S wave in lead V_6.

Right ventricular hypertrophy can be diagnosed by either of the following criteria: (a) the R wave in lead V_1 is greater than normal for age or (b) the S wave in lead V_6 is greater than normal for age.

A positive T wave in lead V_1 in patients between the ages of 7 days and 10 years supports the diagnosis of right ventricular hypertrophy.

RVH/RVE criteria:
R in V_1 > normal for age
S in V_6 > normal for age
rSR' in V_1 with R' > R and R' > 5 mm
Upright T wave in V_1 between age 1 week and 12 years
RAD (right axis deviation of QRS frontal plane axis)

Differentiating RVH and RVE:
Patterns reflecting increases in right ventricular muscle mass ("hypertrophy") usually show a tall R wave in lead V_1 whereas patterns showing right ventricular enlargement usually show an rSR' pattern in lead V_1 and a qRs complex in lead V_6 with a large broad S wave. Usually, the R' exceeds 10 mm. This distinction is not absolute; variations occur.

Left ventricular hypertrophy. The major QRS forces are directed leftward and, at times, posteriorly. Left ventricular hypertrophy can be diagnosed by this "rule of thumb": (a) an R wave in lead V_6 > 25 mm (or >20 mm in children less than 6 months of age) and/or (b) an S wave in lead V_1 > 25 mm (or >20 mm in children less than 6 months of age) (Fig. 1.8).

Combined with ST segment changes and inversion of the T wave in lead V_6, this is referred to as a pattern of "strain" and may be seen in severe left ventricular outflow obstruction.

Distinction between left ventricular hypertrophy and left ventricular enlargement is difficult. Left ventricular hypertrophy may show a deep S wave in lead V_1 and a normal amplitude R wave in lead V_6, whereas left ventricular enlargement shows a tall R wave in lead V_6 associated with a deep Q wave and a tall T wave.
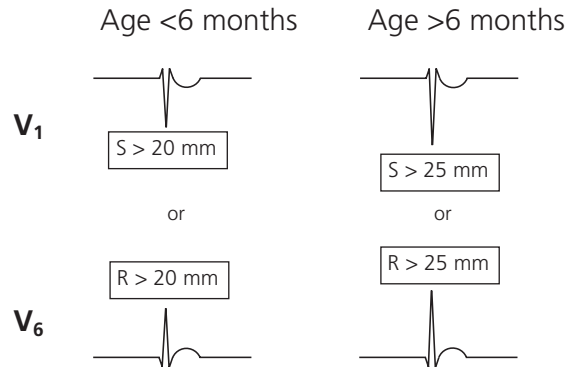
Age <6 months   Age >6 months

Figure 1.8 Electrocardiographic criteria for LVH/LVE by "rule of thumb."

Biventricular hypertrophy. This condition is diagnosed by criteria for both right and left ventricular hypertrophy or by the presence of large equiphasic R and S waves in the mid-precordial leads with a combined amplitude ≥70 mm (Katz–Wachtel phenomenon).

The electrocardiographic standards presented are merely guidelines for interpretation. The electrocardiograms of a few normal patients may be interpreted as ventricular hypertrophy; and indeed, with utilization of these standards only, the electrocardiograms of some patients with heart disease and anatomic hypertrophy may not be considered abnormal.

(3) *QRS duration*. The width of the QRS complex should be measured in lead V_1. The normal range is from 60 to 100 milliseconds; however, infants show shorter QRS intervals. If the QRS complex is greater than 100 milliseconds, a conduction abnormality of ventricular depolarization, such as right or left bundle branch block, is most likely present. In complete right bundle branch block, an rsR′ pattern appears in lead V_1, and the R′ is wide. In lead V_6, the S wave is frequently broad and deep. Right bundle branch block frequently results from operative repair of tetralogy of Fallot. Another example of prolonged QRS duration is Wolff–Parkinson–White syndrome.

Q wave. The Q waves should be carefully analyzed; abnormal Q waves may be present in patients with myocardial infarction. Normally, the Q wave represents primarily depolarization of the interventricular septum. It can be exaggerated if infarction of the left ventricular free wall exists. After the initial 20 milliseconds of the ventricular depolarization, the left ventricular free wall begins depolarizing. With left ventricular infarction, the right ventricular depolarization is

unopposed and directed rightward. This creates a larger and longer Q wave in the left-side leads.

Q wave amplitude. Except in leads aVR, aVL, and V_1, the Q wave should not exceed 25% of the combined amplitude of the QRS complex. If it is larger, the initial QRS forces are accentuated, usually a result of either left ventricular myocardial damage or abnormal septal hypertrophy.

Q wave duration. The Q in leads I, II, and V_6 should be less than 30 milliseconds. If the Q wave duration is longer, myocardial infarction is suspected.

ST segment. The QRS complex returns to the baseline before forming the T wave. The segment (ST) between the QRS complex and the T wave should be isoelectric; but in normal children, particularly adolescents, it may be elevated 1 mm in the limb leads and 2 mm in the mid-precordial leads. It should not be depressed more than 1 mm.

Alterations in the ST segment beyond these limits occur because of myocardial ischemia (depression), pericarditis (elevation), or digoxin (coving depression). The ST segment and T wave are often considered as a unit but should be analyzed separately. ST–T abnormalities are not specific as they can occur in many conditions (e.g., electrolyte disturbances) or in normal children (so-called early depolarization).

T wave. The T wave represents repolarization of the ventricles. Whereas ventricular depolarization takes place from the endocardium to the epicardium, repolarization is considered to occur in the opposite direction. Thus, the direction of the T wave axis is generally that of the QRS axis.

T wave axis. The T wave axis in the frontal plane is normally between $+15°$ and $+75°$; in the horizontal plane, it is between $-15°$ and $+75°$ (Fig. 1.7). In neonates, it begins closer to $-15°$ and moves gradually toward $+75°$ during childhood. Thus in the horizontal plane, the T wave should always be positive in lead V_6. In V_1, the T wave is upright in the first 3 days of life and then becomes inverted until 10–12 years of age, when it again changes to positive.

When both the T wave and the QRS complex, showing either hypertrophy or conduction abnormalities, are abnormal, the T wave abnormalities are probably secondary to the QRS changes.

If, however, the T wave is abnormal while the QRS complex remains normal, the T wave changes represent primary repolarization abnormalities. These may be caused by a variety of factors, such as electrolyte abnormality, metabolic abnormality, pericardial changes, or medication effect.

T wave amplitude. There are no rigid criteria for the amplitude of T waves, although the general rule is the greater the amplitude of the QRS, the greater the T wave. The average T wave amplitude is approximately 20% of the average QRS amplitude. T waves normally range from 1 to 5 mm in standard leads and from 2 to 8 mm in precordial leads.

T wave amplitude is affected by serum potassium concentration. Hypokalemia is associated with low voltage T waves, and hyperkalemia is associated with tall, peaked, and symmetrical T waves. A variety of T wave patterns have been associated with other electrolyte abnormalities.

T wave duration. Best measured by the QT interval, defined as the time from onset of the Q wave to termination of the T wave, it varies naturally with heart rate. Therefore, it needs to be corrected for heart rate by measuring the interval between R waves (R–R). Thus, the equation representing this follows:

$$QT_c = \frac{QT}{\sqrt{R-R}},$$

where QT_c is corrected QT interval (seconds); QT is measured QT interval (seconds); and R–R is measured interval between R waves (seconds).

Males:

normal $QT_c \leq 440$ milliseconds

Females:

normal $QT_c \leq 450$ milliseconds

The QT_c normally does not exceed 440 milliseconds for males and 450 milliseconds for females. Hypercalcemia and digitalis shorten the QT_c; hypocalcemia lengthens it. Other medications may variably affect the QT_c.

Long QT syndrome (LQTS) is a familial condition associated with syncope, seizures, ventricular tachycardia, and sudden death.

U wave. In some patients, a small deflection of unknown origin, the U wave, follows the T wave. It can be prominent in patients with hypokalemia or hypothermia.

Chest radiography

Chest X-rays should be considered for every patient suspected of cardiac disease. Study of the X-ray films reveals information about cardiac size, the size of specific cardiac chambers, the status of the pulmonary vasculature, and the

variations of cardiac contour, vessel position, and organ situs. Two views of the heart are usually obtained, posteroanterior and lateral.

Cardiac size

Size can be evaluated best on a posteroanterior projection.

> Cardiac enlargement indicates an augmented volume of blood in the heart. Any condition that places a volume load upon the heart (e.g., an insufficient valve or a left-to-right shunt) leads to cardiac enlargement proportional to the amount of volume overload.
>
> In contrast, ventricular hypertrophy, meaning increased thickness of the myocardium, does not show as cardiac enlargement on the roentgenogram, although it might change the contour of the heart.

Care must be taken in interpreting X-rays of neonates, particularly those obtained in intensive care units with portable equipment. Three factors in this situation can result in an image that falsely appears as cardiomegaly. The films are usually obtained in anteroposterior rather than posteroanterior projection; the X-ray source-to-film distance is short (40 inches rather than the standard 72 inches); and the infant is supine. (In all supine individuals, cardiac volume is greater.)

The anatomic position of the cardiac chambers in the roentgenographic views is shown in Fig. 1.9. Several important anatomic features are illustrated. The atria and ventricles, rather than being positioned in true right-to-left relationship, have a more anteroposterior orientation. The right atrium and right ventricle are anterior and to the right of the respective left-sided chambers. The interatrial and interventricular septae are not positioned perpendicular to the anterior chest wall but at a 45° angle to the left and 35% tilted away from the midline of the body.

In the posteroanterior projection, the right cardiac border is formed by the right atrium. Prominence of this cardiac border may suggest right atrial enlargement, but this diagnosis is difficult to make from the roentgenogram.

The left cardiac border is composed of three segments: the aortic knob, pulmonary trunk, and broad sweep of the left ventricle. The right ventricle does not contribute to the left cardiac border in this projection.

Prominence of the aorta or the pulmonary trunk may be found on this view. Enlargement of either of these vessels occurs in three hemodynamic situations: increased blood flow through the great vessel, poststenotic dilation, or increased pressure beyond the valve, as in pulmonary hypertension. A concave pulmonary arterial segment suggests pulmonary artery atresia or hypoplasia and diminished volume of pulmonary blood flow.
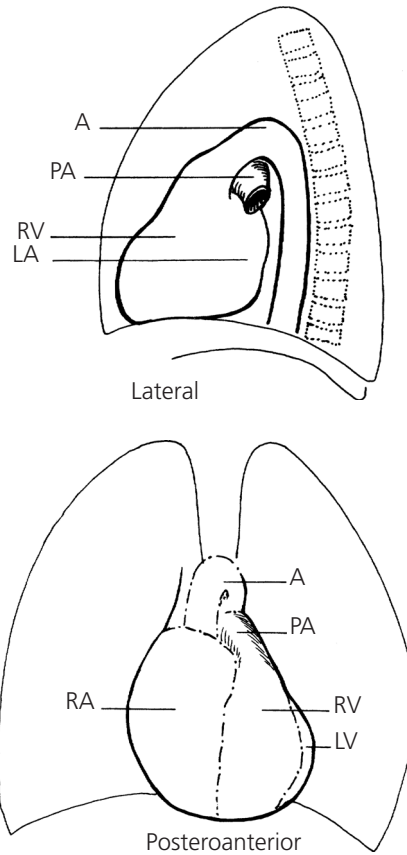
46   Pediatric cardiology

Figure 1.9 Relationship of cardiac chambers observed in posteroanterior and lateral chest X-rays. (Abbreviations: A, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle.)

On the lateral film the margins of the cardiac silhouette are formed anteriorly by the right ventricle and posteriorly by the left atrium. This view is preferred for showing left atrial enlargement because the left atrium is the only cardiac chamber that normally touches the esophagus. A barium swallow can be employed to delineate the esophagus. In a normal individual, the left atrium may indent the anterior esophageal wall, but the posterior wall is not displaced.

If both anterior and posterior walls are displaced, left atrial enlargement is present.

Normally, the lower part of the right ventricle abuts the sternum and air-filled lung extends down between the sternum and the right ventricle and pulmonary artery. When the latter retrosternal space is obliterated by cardiac density, right ventricular enlargement is present; however, in infants this space may also be obliterated by the thymus.

> Both the electrocardiogram and the chest X-ray may be used to assess cardiac chamber size. Left atrial enlargement is best detected by chest X-ray, whereas ventricular or right atrial enlargement is better detected by electrocardiogram.

Cardiac contour

In addition to the search for information about cardiac size on the posteroanterior view of the heart, the practitioner should direct attention to distinctive cardiac contours, such as the boot-shaped heart of tetralogy of Fallot. In conditions with right ventricular hypertrophy, the cardiac apex may be turned upward, whereas conditions with left ventricular hypertrophy or dilation lead to displacement of the cardiac apex outward and downward toward the diaphragm.

Situs

Note situs of the heart, stomach, and especially the aortic arch. In infants with a prominent thymus, the aortic knob is usually obscured, and normal aortic arch position is inferred from the rightward displacement of the trachea in a properly positioned posteroanterior chest film. A right aortic arch is common in tetralogy of Fallot and truncus arteriosus and can be diagnosed by leftward displacement of the trachea.

Pulmonary vasculature

The status of the pulmonary vasculature is the most important diagnostic information derived from the chest X-ray; this function has not been replaced by the echocardiogram. The radiographic appearance of the lung blood vessels reflects the degree of pulmonary blood flow. Because many cardiac anomalies alter pulmonary blood flow, proper interpretation of pulmonary vascular markings is diagnostically helpful. It is one of the two major features discussed in this book for initiating the differential diagnosis.

The lung fields are assessed to determine if the vascularity is increased, normal, or diminished, reflecting augmented, normal, or decreased pulmonary blood flow respectively. As a check of the logic of interpretation, the vascular

markings should be compared with cardiac size. If a large volume left-to-right shunt exists, the heart size has to be larger than normal.

Pulmonary vascular markings may be more difficult to analyze from portable films obtained in a neonatal care unit because X-ray exposure time is longer, resulting in blurred images from rapid respirations, and from the redistributed pulmonary blood volume in the supine patient.

With experience obtained from viewing a number of chest X-rays, the status of pulmonary vasculature can be judged. With increased vascularity, the lung fields show increased pulmonary arterial markings; the hilae are plump; and vascular shadows radiate toward the periphery. With decreased vascularity, the lungs appear dark or lucent; the hilum is small; and the pulmonary arterial vessels are stringy.

Summary of chest X-ray parameters
  Situs (heart, stomach, and aortic arch)
  Cardiac size
  Cardiothymic silhouette, shape, and contour
  Pulmonary artery silhouette
  Pulmonary vascular markings (normal, increased, or decreased;
    symmetric versus asymmetric)

Pulse oximetry

Because oxyhemoglobin and deoxyhemoglobin absorb light differently, spectrophotometry can be used to measure the percent of hemoglobin bound to oxygen.

Pulse oximeters utilize a light source and light sensor applied to the surface of a patient's skin to noninvasively compare the light absorption of moving blood (during arterial flow) with the light absorption of the nonmoving blood and tissue during arterial diastole (analogous to a reference sample).

Functional arterial oxygen saturation (SaO_2) in percent is calculated automatically and displayed along with pulse rate.

Pulse oximeters do not detect dysfunctional hemoglobin (e.g., methemoglobin and carboxyhemoglobin), so patients with important concentrations of these types of abnormal hemoglobin have a factitiously high SaO_2 compared with their true fractional saturation as measured from a blood sample using a standard laboratory co-oximeter.

Other factors affecting pulse oximeter results include skin pigmentation, poor skin perfusion, tachycardia, ambient light, and shifts in the oxyhemoglobin absorption spectrum that can accompany chronic cyanosis.

Neonates with cyanotic heart malformations (e.g., transposition of the great vessels) or obstructive lesions (e.g., coarctation of the aorta) may have differential cyanosis, a measurable inequality in the pulse oximetry readings from

preductal (right hand) compared to postductal (foot) sites, even though the difference is not apparent by physical examination.

Blood counts

In infants and children with cyanotic forms of congenital cardiac disease, hypoxemia stimulates the bone marrow to produce more red blood cells (polycythemia), thus improving oxygen-carrying capacity. As a result, both the total number of erythrocytes and the hematocrit are elevated. The production of the increased red cell mass should be paralleled by an increase in hemoglobin. In a patient with cyanosis and normal iron stores, the hemoglobin should be elevated and the red cell indices normal.

Iron deficiency

In infancy, however, iron deficiency is common; it may be accentuated in cyanotic infants because of the increased iron requirements and by the fact that such infants may have poor appetites and a milk diet primarily. In such infants, the red cell indices reflect iron deficiency anemia because the hemoglobin value is low relative to the red blood count and the hematocrit. In fact, a cyanotic infant may have a hemoglobin value that is normal or even elevated for age and still suffer from iron deficiency.

> An example is an infant with a hemoglobin of 16 g/dL and a hematocrit of 66%. The hematocrit value reflects the volume of red cells elevated in response to hypoxemia; the hemoglobin value primarily reflects the amount of iron available for its formation. In this infant the hemoglobin should be 22 g/dL. (Normally, the number for the hemoglobin value should be one third that of the number of the hematocrit value.)

The mean corpuscular volume is always low in iron deficiency, even if the hemoglobin is normal or above normal. An iron-deficient infant may improve symptomatically following administration of iron. Iron deficiency has been associated with an increased risk of stroke in severely polycythemic patients.

Patients with inoperable cyanotic heart disease should have hemoglobin and hematocrit values periodically measured; discrepancies between the two should be noted and managed. Similar information may be obtained by evaluating a blood smear. Serum iron testing is usually unnecessary.

Hyperviscosity

Although vascular resistance varies with viscosity, which itself varies with polycythemia, the effect is not clinically important until the hematocrit approaches 70%. In general, adolescents and young adults with inoperable cyanotic heart disease worsen with phlebotomy, probably because of its detrimental effects

on oxygen-carrying capacity. Iron deficiency usually complicates repeated phlebotomy.

Anemia

Anemia may increase the cardiac workload in patients with congestive heart failure and may predispose tetralogy patients to have hypercyanotic spells. In cyanotic patients, severe anemia may lead to an important decrease in the oxygen-carrying capacity.

Echocardiography

Echocardiography, a powerful noninvasive diagnostic technique, requires a high degree of skill in performance and interpretation. This method adds considerable information regarding cardiac function and structure to that gained from history, examination, electrocardiogram, and chest radiography.

Echocardiography of pediatric patients is considerably different from that of adults, both in the technical performance required to obtain quality information in sometimes less than cooperative children and in the interpretation, which emphasizes anatomic relationships, connections, and physiologic principles more than the mere recording of chamber size and ventricular function. Because of poor acoustic penetration, the latter information is often the most that can be obtained from an adult patient without resorting to more invasive techniques, like transesophageal echo, in which the heart is imaged from a probe positioned in the esophagus instead of on the anterior chest wall. In most pediatric patients, excellent images are obtained using surface (transthoracic, or TTE) echo alone so that other techniques, such as transesophageal echo (TEE), are reserved for special circumstances (e.g., intraoperative echo) where surface images would be impossible to obtain. Infants and children are not routinely sedated for echocardiography since a complete and high-quality echocardiogram can usually be done without sedation.

Echocardiography is based on a familiar principle illustrated by bats, which emit ultrahigh frequency sound waves that are reflected from surfaces and are received back, allowing the bats to judge their surroundings and to avoid collision with objects. The principles of Doppler determination of the velocity of moving objects is applied to determine the speed and direction of blood flow.

Two-dimensional images

An echocardiogram is recorded by placing a transducer in an interspace adjacent to the left sternal border and at other locations on the chest and abdomen (Fig. 1.10). The small transducer contains a piezoelectric crystal that converts electrical energy to high-frequency sound waves. Thus, the transducer emits sound waves into the chest that strike cardiac structures; these sound waves (echoes) are then reflected back to the chest wall. The transducer
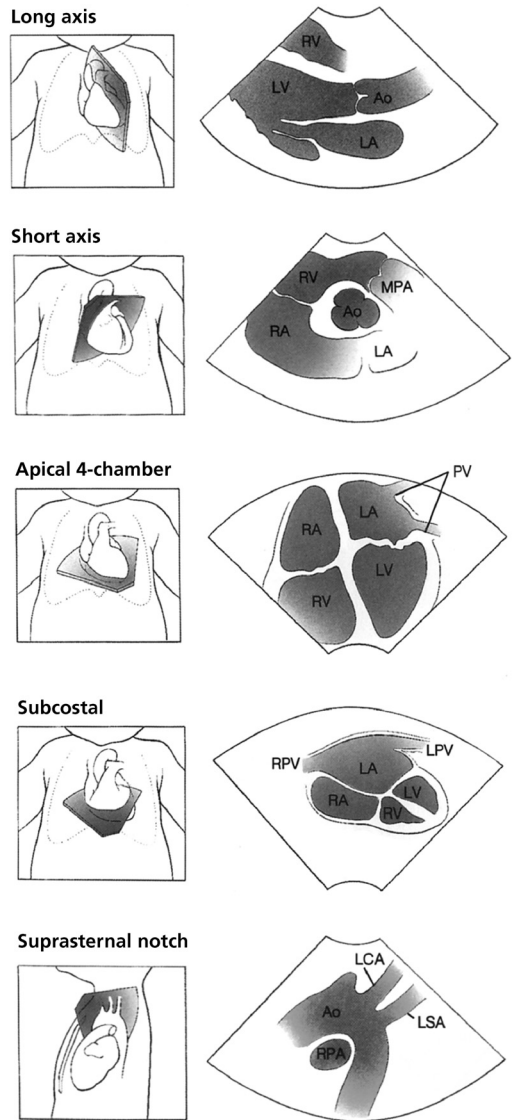
Long axis

Short axis

Apical 4-chamber

Subcostal

Suprasternal notch

Figure 1.10 Two-dimensional (2D) echocardiography. Five standard views are shown. The left-sided illustrations show the sector-shaped plane of the ultrasound beam inscribed on the patient's chest; the right-sided illustrations show the corresponding 2D images of the heart and vessels. (Images courtesy of Agilent Technologies.)

52 Pediatric cardiology

receives sound (echoes) from the cardiac structures and reconverts them to electrical energy that is then recorded.

Because the frequency of the sound waves and the speed of sound in body tissues are constant, the interval between the emission of sound and the return of sound indicates the distance into the heart and back that the sound wave traveled. The ultrahigh frequency sounds are reflected only from interfaces between structures of different density, such as the interface between the ventricular cavity (blood) and the ventricular septum (muscle). The amount of sound returned depends on the nature of the substances on either side of the interface.

The reflecting surface must be perpendicular to the transducer; when a surface lies tangential, the sound waves are generally reflected in a different direction and are not received by the transducer. As the sound waves travel into the heart, at each interface some sound returns to the transducer, and some continues to the next structure where more is reflected, while some still continues. In this way, multiple sound waves are reflected at various distances from the surface of the chest; these echoes are used to generate two-dimensional images moving in real time.

M-mode

In M (movement) mode (Fig. 1.11), the vertical axis represents distance from the transducer on the surface of the chest and the horizontal axis represents time. The movements of cardiac structures can be recorded over several cardiac cycles. A simultaneous electrocardiogram assists in the timing of cardiac events.
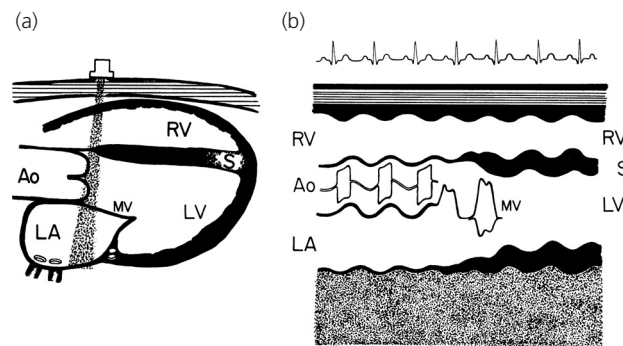

Figure 1.11 M-mode echocardiogram and two-dimensional (2D or cross-sectional) echocardiogram compared. The transducer beam passing through the cross-sectional view (a) corresponds to the same structures seen in the M-mode (b) during a "sweep" of the transducer from aorta to ventricles. (Abbreviations: Ao, aorta; LA, left atrium; LV, left ventricle; MV, mitral valve; RV, right ventricle; S, interventricular septum.)

Table 1.8 Echocardiographic Upper Limits of Left Ventricular (LV) Dimensions by Body Weight.

Body Weight (kg)	LV Diastolic Diameter (cm)	LV Diastolic Wall Thickness (mm)
4	2.5	5
8	3.0	6
15	3.5	7
30	4.5	9
60	5.0	10

Courtesy William S. McMahon, MD, based on published data, including Henry WL, Ware J, Gardin JM, et al. Echocardiographic measurements in normal subjects: Growth-related changes that occur between infancy and early adulthood. Circulation 1978;57:278–285.

Chamber size and wall thickness are usually measured by M-mode. Representative normal left heart values are shown in Table 1.8.

Cardiac function is estimated by M-mode. Although not true measures of contractility, left ventricular shortening fraction (percent change in diameter between diastole and systole; normal $\geq 28\%$) and ejection fraction (percent change in estimated volume; normal $\geq 55\%$) are often both used to describe systolic ventricular function. These values may vary with changes in afterload, preload, or contractility.

Calculation of Shortening Fraction (SF) and Ejection Fraction (EF)

$$SF\,(\%) = \frac{LVEDD - LVESD}{LVEDD} \times 100$$

$$EF\,(\%) \cong \frac{LVEDD^3 - LVESD^3}{LVEDD^3} \times 100$$

or

$$EF \cong SF \times 1.7$$

normal

$SF \geq 28\%$

$EF \geq 55\%$

LVEDD left ventricular end-diastolic diameter
LVESD left ventricular end-systolic diameter

Doppler

Doppler echocardiography provides information on the direction and speed (velocity) of moving blood. Three main types of Doppler echocardiography are commonly used.

Pulsed wave Doppler. Pulsed wave (PW) Doppler derives velocity information from discrete packets of ultrasound transmitted and then received by the transducer, allowing precise interrogation of small regions of a vessel or chamber. The main limitation of PW Doppler is the compromise between the depth of the structure to be Dopplered and the maximum velocity that can be measured—maximum velocity decreases as the distance to the target increases.

Continuous wave Doppler. Continuous wave (CW) Doppler uses simultaneous continuous transmission and receipt of ultrasound and provides highly accurate estimates of very-high velocity blood flow—for example, through a stenotic aortic valve—but cannot localize the source of the fastest velocities, as PW Doppler can.

Both PW and CW Doppler are commonly used to determine the following:

(1) *Pressure gradient*. Just as river water speeds up passing through a narrow rapids, Doppler velocities can be used to predict pressure gradients between two chambers according to a simplified form of the Bernoulli equation, given a constant flow rate.

$$PG = V^2 \times 4,$$

where PG is pressure gradient (millimeters of mercury; mm Hg); V is velocity (meters/second; m/s) of blood flow; and 4 is a constant.

This technique is commonly used to estimate the pressure gradient across a stenotic valve, such as aortic stenosis.

Also, the maximum velocity of blood regurgitating through an AV valve during systole (depending on atrial pressure) gives an approximation of the peak systolic pressure in the ventricle.

(2) *Flow (cardiac output)*. In areas where flow is laminar (most of the blood is moving at the same velocity at any given point in time), Doppler can be used to measure the change in this velocity throughout the systolic ejection period. The mean velocity (cm/s) during ejection through a normal semilunar valve of known area (cm^2) can be used to calculate the flow (cm^3/s of ejection) and combined with the heart rate to determine cardiac output (cm^3/s, or L/min).

Color (flow velocity mapping) Doppler. Color Doppler allows the generation of a color-coded display of real-time blood flow velocity and direction overlaid on the black-and-white two-dimensional image of the heart. Color Doppler allows visualization of jets of blood flow, such as in a small VSD or

for grading the degree of insufficiency of a valve. Physiologic blood flow is easily demonstrated with color Doppler: by convention, flow away from the transducer is represented by blue and flow toward the transducer is red. The colors have no relationship to blood oxygen levels.

Specialized echocardiography

Transesophageal echocardiography. Both TTE and TEE are important diagnostic techniques in children. In general, in infants and children, the range of structures that can be evaluated is greater with TTE and the image quality comparable to TEE. For patients undergoing cardiac surgery or catheterization, TEE is often employed concurrently. TEE usually requires sedation and/or anesthesia whereas many centers do not routinely sedate children for TTE. The size of the available transesophageal transducer may limit the technique to larger infants.

Intracardiac echocardiography. Intracardiac echocardiography (ICE) utilizes a catheter-mounted transducer to acquire intravascular and intracardiac image and Doppler data during cardiac catheterization, usually electrophysiologic catheterization, and provides more precise localization of structures than fluoroscopy and angiography.

Tissue Doppler imaging. Tissue Doppler imaging, done at the time of TTE or TEE, uses Doppler principles to measure the velocity of the ventricular walls, rather than the movement of blood, as in standard Doppler, providing information about ventricular performance and regional wall motion abnormalities.

Three-dimensional echocardiography. Three-dimensional echocardiography (3D echo) generates a real-time pseudo-holographic representation of the heart using a "stack" of sequential 2D images, allowing enhanced images of complex structures such as AV valves and ventricular outflow tracts.

Magnetic resonance imaging (MRI and MRA)

This technique generates high-quality static images of the body that are similar to those of computed tomography except that ionizing radiation is not used. Rather, a powerful magnetic field surrounds the patient, and the chest is irradiated with radiofrequency pulses from nearby coils that produce alignment of the normally random arrangement of the atomic nuclei of paramagnetic elements. Since hydrogen in water and fat is the most common atom in the body, most images are created using the radiofrequency emitted from these hydrogen nuclei and received as induced current in surrounding coils.

A basic assumption of MRI is that the subject is stationary, a problem partly overcome during cardiac imaging by gating the acquisition of signals to respirations and the electrocardiogram.

Although multiple images can be acquired and "cinelooped" to create the illusion of movement, considerable time is required to create each image, so "real-time" images, such as those obtained with echocardiography, are not possible (see Table 1.9). Since the patient must lie still for the acquisition of all the multiple images, sedation is required for infants and small children, which, although MRI is noninvasive and involves no radiation, increases the relative risk of the procedure.

MRI can provide some data regarding pressure gradients, but the speed and ease of acquisition are not comparable to Doppler echocardiography. MRI does provide excellent images in large adolescents and adults where echocardiography is impossible, but patients with certain magnetic implants, such as artificial pacemakers and certain prosthetic devices, cannot be subjected to the intense magnetic field required. Intravenous nonionic contrast agents are often employed, especially with magnetic resonance arteriography.

Computed tomography

Computed tomography (CT) for cardiovascular imaging has many of the same advantages and disadvantages of MRI and MRA. Computed tomographic angiography (CTA) utilizes higher resolution, faster CT instruments, along with the intravenous administration of iodinated contrast, to obtain very high-quality images; however, the normally faster heart rates of children limit resolution and hemodynamic data is limited. CTA gated to the patient's electrocardiogram provides higher resolution images of moving cardiac structures, yet result in significantly higher radiation doses. See Table 1.9 comparing various imaging techniques.

Exercise testing

This technique is helpful in several situations but requires the cooperation of the child. Hence very young children are not candidates for testing. The authors usually limit exercise testing to children over the age of 6 years. Dobutamine challenge has been used as an alternative, with assessment of myocardial performance by echocardiography and myocardial perfusion using nuclear scans.

Indications

Pre- and postoperative assessment. Preoperative assessment of obstructive lesions (e.g., aortic stenosis) may benefit patients with borderline gradients because it helps decide the timing of intervention. Many patients have indications for intervention (surgery or catheterization) independent of exercise data.

Postoperative assessment. Postoperative assessment of cardiopulmonary function (using maximal oxygen consumption and/or exercise endurance time) helps in symptomatic patients and in those with mild systolic dysfunction. It can also aid in formulating sports or occupational recommendations for adolescents and adults with congenital heart disease.

Table 1.9 Comparison of Common Diagnostic Imaging Modalties in the Evaluation of Congenital Heart Disease Patients.

	CXR	Ba Eso	CT	CTA	MRI/MRA	Echo TTE	Echo TEE	Cath
Realtime	N	Y/N	N	N	N	Y	Y	Y
Hemodynamics	–	–	–	+	+	+++	++	++++
Availability	++++	+++	+++	++	+	++	+	+
Interpretation	+	++	+++	+++	++++	++++	++++	++++
Cost	+	++	+++	++++	++++	++	++	++++
Radiation	+	++	+++	++++	–	–	–	++++
Anesthesia and/or sedation	N	N	Y/N	Y/N	Y/N	N/Y	Y	Y
IV contrast	N	N	N/Y	Y	Y	N	N	Y
Heart rate effect	–	–	++	+++	++	–	–	–
Respiratory and movement effect	+	–	+	+	++	–	–	–
Most useful data/condition	Pulmonary blood flow, degree and symmetry	Vascular ring/sling	Rapid assessment aortic dissection; effusions	Detailed images. Short acquisition time	Detailed images; best for static structures	Realtime, chamber dimension, thickness, hemodynamics, anatomic relationships and situs	Enhanced images when TTE suboptimal, or intraop, intracath	Intervention. Diagnosis of PA htn and reactivity
Disadvantages	No direct hemodynamic data	No direct imaging of anomaly	Relatively low resolution	Heart rate, resp artifact	Heart rate, resp artifact	Acoustic windows become more limiting as pt size increases	Some structures imaged by TTE not seen by TEE	Radiation and contrast load. Vascular entry sites required

CXR, Chest radiograph; Ba Eso, Barium Esophagogram (table refers to fluoroscopically-performed examinations; simple studies can be accomplished with barium swallow at the time of an upright CXR in some patients); CT, computed tomography of the chest; CTA, computed tomographic, angiography; requires higher-resolution equipment than standard CT; MRI/MRA, magnetic resonance imaging/magnetic resonance arteriography; Echo TTE, echocardiogram, transthoracic; Echo TEE, echocardiogram, transesophageal; Cath, cardiac catheterization.

Myocardial ischemic syndromes. Suspected coronary artery insufficiency (e.g., Kawasaki disease with aneurysm or stenosis or postoperative anomalous coronary artery origin repair) is most sensitively assessed by a combination of electrocardiographic and nuclear perfusion studies done during a maximal exercise study. Exercise electrocardiography alone has a false-negative rate of 15% in adults.

Arrhythmias

Wolff–Parkinson–White syndrome. Patients with this condition may be at greater risk for life-threatening ventricular tachyarrhythmia if the delta wave persists at sinus rates of >180 bpm.

Premature ventricular contractions. If benign, these usually disappear at fast sinus rates during exercise.

Atrioventricular block. The rate reserve of the patient's natural subsidiary (backup) pacemaker can be assessed during exercise.

Suspected long QT syndrome. Patients with this condition do not show the usual shortening of the QT interval as the heart rate increases.

Tachyarrhythmia. Patients with documented tachyarrhythmia (SVT or VT) during exercise or those at risk during exercise (e.g., postoperative tetralogy of Fallot) may be candidates for drug treatment; exercise assesses the efficacy of the treatment.

Patients with a history of palpitations only usually have normal exercise tests and are better studied using outpatient electrocardiographic monitoring to document the rhythm during symptoms.

Syncope. Usually, only patients with a history of syncope during exercise need study.

Hypertension. Postoperative coarctation patients and some with other forms of systemic hypertension may register as normotensive (or borderline) at rest but may exhibit an exaggerated systolic hypertensive response to exercise.

Procedure

Specialized equipment is used for grading the workload and for continuously recording multilead electrocardiograms.

> Heart rate rises linearly to an age-related maximum (200–210 bpm for normal children and adolescents).
>
> Systolic blood pressure rises to a normal maximum of 180–215 mm Hg, whereas diastolic pressure remains constant or falls slightly.

If indicated, pulse oximetry and oxygen consumption are measured.

Stress echocardiography allows determination of cardiac function or change in gradients but can be technically challenging.

Spirometry before and after exercise is useful if exercise-induced bronchospasm is suspected.

A bicycle ergometer allows more precise setting of the workload but is often limited to larger patients. A treadmill is more common. The Bruce protocol involves increasing treadmill speed and inclination in stages every 3 minutes; because smaller children are unable to run at the maximum speed (6 mph) of the Bruce protocol, most pediatric laboratories use the modified Bruce protocol, which limits the speed to a maximum of 3.4 mph.

Risks
Risks of syncope, arrhythmia requiring immediate treatment, or death are higher in certain conditions, including hypertrophic cardiomyopathy, pulmonary vascular obstructive disease, severe aortic stenosis, uncontrolled hypertension, and severe dilated cardiomyopathy. The potential benefits of exercise testing may not warrant the risk in many of these patients.

Cardiac catheterization
Cardiac catheterization requires a staff of trained specialists—pediatric cardiologists, radiologists, laboratory technicians, and nurses. As a diagnostic procedure it provides detailed information about the heart not found by other techniques. Its use as a diagnostic technique has decreased as its application for treatment (intervention) has expanded.

Diagnostic cardiac catheterization
Cardiac catheterization was used until recently to establish the diagnosis in most children with cardiac malformation. Now, with the use of echocardiography and other noninvasive studies, the indications for diagnostic cardiac catheterization have become targeted: to acquire specific anatomic (e.g., coronary artery anatomy in transposition), functional (e.g., pulmonary vascular resistance in an older child with a VSD), or histologic (cardiac biopsy in a transplant patient) information.

Interventional therapeutic catheterization

Interventional catheterization began in the 1960s with the Rashkind atrial septostomy, which used a spherical latex balloon pulled forcefully through a patent foramen ovale to create a large ASD for palliation of d-transposition of the great vessels.

Currently, radial balloon dilation with sausage-shaped catheter-mounted balloons is commonly used to relieve obstruction in stenotic semilunar valves and nonvalved pathways (e.g., recurrent coarctation, stenotic pulmonary arteries).

Catheter-based methods for closure of patent ductus arteriosus and ASD are used widely, and devices for closure of certain VSDs are available.

Electrophysiologic catheterization

Electrophysiologic catheterization is performed to define the mechanism and characteristics of arrhythmias.

Radiofrequency ablation or cryoablation may be used to eliminate accessory electrical connections or automatic foci, thus curing certain arrhythmias.

Procedure

Cardiac catheterization is performed in children in a manner that ensures a quiet, controlled, and safe environment for the child; allows minimum discomfort, pain, and anxiety; and also achieves optimum data collection or treatment.

Anesthesia. Two basic approaches are used.

General anesthesia. Anesthesia, usually with endotracheal intubation in neonates, infants, and small children, allows precise control of airway and ventilation. This avoids pulmonary vascular resistance elevation that may accompany hypoventilation from oversedation.

Sedation. Sedation alone is successfully used in patients of all ages at many centers. This usually involves a combination of agents, including narcotics, benzodiazepines, phenothiazines, and ketamine.

Vascular access. Both the right and left sides of the heart may be catheterized either by percutaneous puncture (Seldinger technique) or by operative exposure ("cutdown") to major peripheral veins and arteries. The right side of the heart is accessed through veins in the inguinal area or upper body (e.g., internal jugular vein). The left side of the heart can be catheterized through two approaches: a venous catheter may be passed through the foramen ovale or ASD (or via a tiny defect created with a needle-tipped catheter) into the left atrium or an arterial catheter may be inserted into the brachial or femoral artery and passed retrograde across the aortic valve into the left ventricle. Arterial puncture and atrial septum puncture carry more risk than do venous studies.

Table 1.10 Normal Cardiac Catheterization Values.

Site	Oxygen Saturation (%)	Pressure (mm Hg)
RA	70 ± 5	mean 3–7
RV	70 ± 5	25/EDP 0–5
PA	70 ± 5	25/10, mean 15
LA, PCW	97 ± 3	mean 5–10
LV	97 ± 3	100/EDP 0–10
Aorta	97 ± 3	100/70, mean 85

EDP, end-diastolic pressure; LA, left atrium; LV, left ventricle; PA, pulmonary artery; PCW, pulmonary capillary wedge pressure; RA, right atrium; RV, right ventricle.

Technique. Once the catheter has been inserted into the vessel, it can be advanced into the heart and directed into various cardiac chambers and major blood vessels with the aid of fluoroscopy. At any of these sites, pressures can be measured, blood samples obtained, and contrast media injected.

Pressure data. The catheter is connected to a pressure transducer and the values obtained are compared with normal (Table 1.10) to evaluate stenotic lesions or pulmonary hypertension.

Oximetric data. Blood samples from each cardiac site are analyzed for oxygen content or hemoglobin saturation to determine if a shunt is present. Normally, the oxygen saturation in each right-sided cardiac chamber is similar, but an increase in the oxygen saturation in any chamber, compared with the preceding site, may mean a left-to-right shunt at that level. Normal variations in oxygen content occur, so a slight increase may not indicate a shunt. Multiple samples at each site are used to resolve this point.

Normally, the oxygen saturation of blood in the left atrium, the left ventricle, and the aorta should be at least 94%; if less, a right-to-left shunt is present.

Derived values. The pressure and oximetry data can be used to derive various measures of cardiac function.

Flow (cardiac output). This can be calculated using the Fick principle:

$$\text{Cardiac output (L/min)} = \frac{\text{Oxygen consumption (mL } O_2 \text{ / min)}}{\text{Arteriovenous oxygen difference (mL/dL)} \times 10}$$

The patient's rate of oxygen consumption can be determined by analyzing a timed collection of the patient's expired air.

The arteriovenous oxygen difference is obtained by analyzing blood samples drawn from the arterial side of the circulation (aorta or peripheral artery) and from the venous side of the heart (usually the pulmonary artery). The oxygen content (mL of O_2/dL whole blood) is used, as the percent hemoglobin saturation alone cannot be used in this calculation.

Cardiac output determined by the Fick principle is widely used in analyzing catheterization data and has become the standard to which other methods of determining cardiac output, such as thermodilution, are compared.

Since many cardiac malformations have either a left-to-right or a right-to-left shunt, the blood flow through the lungs may differ from that through the body, even though the oxygen consumption in the body must equal the oxygen picked up in the lungs. The Fick principle may still be used for such patients:

$$Q_S = \frac{\dot{V}_{O_2}}{SA - MV},$$

where Q_S is systemic blood flow (L/min); $\dot{V}_{O_2}$ is oxygen consumption (mL O_2/min); and $SA - MV$ is systemic arterial–mixed venous oxygen difference (mL O_2/L blood).

$$Q_P = \frac{\dot{V}_{O_2}}{PV - PA},$$

where Q_P is pulmonary blood flow (L/min); $\dot{V}_{O_2}$ is oxygen consumption (mL O_2/min); and $PV - PA$ is pulmonary venous–pulmonary arterial oxygen difference (mL O_2/L blood).

Pulmonary/systemic blood flow ratio (Q_P/Q_S). Without assuming or measuring the oxygen consumption, the pulmonary blood flow (Q_P) can be expressed as a ratio of the systemic blood flow (Q_S):

$$\frac{Q_P}{Q_S} = \frac{SA - MV}{PV - PA},$$

where SA, MV, PV, and PA represent oxygen saturations (%).

Except for oxygen saturation (%), all other variables required for oxygen content calculation (e.g., hemoglobin concentration) cancel out of the equation.

Resistance. Systemic and pulmonary vascular resistances can be calculated from the hydraulic equivalent of Ohm's law:

$$R = \frac{P}{Q},$$

where R is resistance; P is mean pressure drop across a vascular bed; and Q is cardiac output.

Table 1.11 Normal Derived Cardiac Catheterization Values.

Cardiac Index (CI)*	3–5 L/min/m^2
Pulmonary resistance (R_P)*	2units • m^2
Systemic resistance (R_S)*	10–20 units • m^2
Resistance ratio (R_P/R_S)	0.05–0.10

*Values indexed to body surface area.

Therefore

$$R_S = \frac{\overline{SA} - \overline{RA}}{Q_S},$$

where R_S is systemic vascular resistance (mm Hg/L/min); $\overline{SA}$ is mean systemic artery (aortic) pressure (mm Hg); $\overline{RA}$ is mean systemic vein (right atrial) pressure (mm Hg); and Q_S is systemic blood flow (L/min).

$$R_P = \frac{\overline{PA} - \overline{LA}}{Q_P},$$

where R_P is pulmonary vascular resistance (mm Hg/L/min); $\overline{PA}$ is mean pulmonary arterial pressure (mm Hg); $\overline{LA}$ is mean pulmonary vein (left atrium or pulmonary capillary wedge) pressure (mm Hg); and Q_P is pulmonary blood flow (L/min).

Resistance ratio (R_P/R_S) can similarly be calculated from the ratio of the mean pressure differences across the pulmonary and systemic beds, divided by the Q_P/Q_S:

$$\frac{R_P}{R_S} = \frac{(\overline{PA} - \overline{SA})(\overline{LA} - \overline{RA})}{Q_P/Q_S}.$$

Normalization of output and resistance. Resistance is normalized to body surface area, either by using cardiac index (CI) expressed as L/min/m^2 in place of cardiac output in the preceding formulas or by multiplying the raw resistance by the patient's body surface area, which yields resistance in mm Hg • min/L • m^2 or Wood units • m^2 (first described by Paul Wood in the 1950s). Resistance is also expressed as dyne • cm/s^5, which can be converted from Wood units by multiplying by 80.

Normal indexed values are shown in Table 1.11.

Angiocardiography. Radio-opaque contrast material can be injected through the catheter into the cardiac chambers with serial X-ray images obtained digitally or on film (cine angiography), often in two projections simultaneously

64 Pediatric cardiology

(biplane). The imaging system can be rotated around the patient so that angulated projections can be obtained to better visualize various structures (axial angiography). Excellent delineation of the cardiac anatomy can be obtained. Satisfactory details may be illustrated by injecting the material into the pulmonary artery and then imaging as the contrast passes through the left side of the heart (levophase).

Complications of cardiac catheterization. As with any procedure, cardiac catheterization may be associated with complications; the benefits from cardiac catheterization must clearly outweigh the risks.

Death. Death is extremely uncommon (<0.1%) in children beyond 1 year of age. The risk is higher in infants, particularly neonates, who are often critically ill and require catheterization so that a lifesaving catheter intervention or operation can be performed.

Vascular complications. Rarely, compromise of blood vessels used for catheter entry occurs. Temporary or permanent occlusion of the femoral vein or entire inferior vena cava may occur, which may cause transient venous stasis and edema in the lower extremities. Seldom dangerous, the major impact is the inability to reenter these vessels if the patient requires additional catheterization.

Femoral artery injury is more serious, as viability of the limb is at risk. Thrombolytic agents and heparin have been used in the acute management of patients with a pulseless extremity after catheterization.

Rarely, an arteriovenous fistula develops between adjacent vessels used for entry and requires surgery.

Arrhythmia. During most cardiac catheterizations, arrhythmias of some type occur, most often premature ventricular contractions; but these rarely compromise the patient because they tend to be transient. Occasionally, AV block that lasts for several hours occurs.

Radiation. The ionizing radiation dose received by most patients has fallen over the years with improved image-intensifier technology, even though procedure times have lengthened for patients having interventional procedures. Short- and long-term complications from radiation are rare.

ADDITIONAL READING

Driscoll DJ. Fundamentals of Pediatric Cardiology. Philadelphia: Lippincott Williams & Wilkins; 2006.

Lock JE, Keane JF, Perry SB. Diagnostic and Interventional Catheterization in Congenital Heart Disease. Boston: Kluwer Academic Publishers; 1999.

Moller JH, Hoffman JIE. (eds.) Pediatric Cardiovascular Medicine. New York: Churchill
  Livingstone; 2000.
Mullins C. Cardiac Catheterization in Congenital Heart Disease: Pediatric and Adult.
  Oxford: Blackwell; 2005.
Park MK, Guntheroth WG. How to Read Pediatric ECGs, 4th ed. St. Louis: Mosby; 2006.
Park MK. Pediatric Cardiology for Practitioners, 5th ed. St. Louis: Mosby; 2007.
Pelech AN. The cardiac murmur: When to refer? Pediatr Clin North Am 1998;45:107–122.