Clinical Diagnosis in Pediatric Cardiology

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Foreword by

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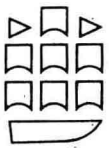

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Foreword

The first major treatises on congenital heart disease in the late 1930s and early 1940s dealt largely with clinical diagnosis based on fluoroscopic and autopsy correlations. Since then the introduction and refinement of cardiac catheterization and selective angiocardiography and, more recently, of echocardiography and radionuclide studies have led to a heavy reliance on laboratory investigation in the diagnosis of heart malformations. A whole generation of physicians, though perhaps unwilling to admit the fact, has come to believe that detailed physical examination as part of the assessment of infants and children with congenital heart disease is to a large extent outmoded and has dubious reliability. For them, physical examination of the cardiovascular system seems relatively unimportant, not worthy of more than a brief period of attention and sometimes carried out for no other reason than to satisfy traditions, shortly about to be buried along with their elderly mentors.

Yet there has never been a time when physical examination better permits recognition, provides quite specific diagnosis very frequently and gives an appreciable distance in predicting the physiologic and anatomic arrangement for a large majority of young patients with heart disease. Ironically this modern bedside and technical capability has derived from the method of the pioneers — a combination of the newer accessory investigations and traditional physical methods.

To abstract key information from the huge reserve of modern clinical facts about congenital heart disease and combine that information into one concise monograph is no mean task but Dr Zuberbuhler has succeeded admirably in doing so. His position as head of one of the largest American services in pediatric cardiology, and his long experience in an environment which has contributed mightily to the subject in hand, give authenticity to the work. For those pediatricians who despair of being able to approach the specific diagnosis of congenital heart disease today amidst the bewildering technology of the 80s, this presentation has particular attraction but all who are involved in the medical supervision of children can benefit from his detailed review.

Toronto, 1981 R.D.R.

Preface

This book was written in the hope that it will be useful to students, house officers and practicing physicians involved in the care of children with heart disease. It is not intended to be either encyclopedic or esoteric; at times complex events have been simplified, perhaps oversimplified, in the interest of brevity and clarity. The descriptions of the clinical findings are largely based on personal experience, with much assistance from colleagues and the literature. Whatever its virtues or faults, this book reflects my conviction that the clinical examination is and will remain an indispensable part of the evaluation of each child with heart disease.

I owe a considerable debt to the members of the cardiology staff at the Children's Hospital of Pittsburgh. Drs Cora Lenox, William Neches and Sang Park reviewed and criticized the manuscript and their suggestions have been invaluable. Dr James Shaver, Chief of Adult Cardiology, University of Pittsburgh, reviewed Chapter 2. These physicians, as well as other members of staff, Drs Robert Mathews, Jay Fricker and Lee Beerman, were kind enough to call the attention of the author to interesting physical findings in patients in our clinic, and many of these were included in the illustrations. Dr Sang Park was especially helpful in the selection and preparation of the illustrations. Ms Virginia Curlee, non-invasive technologist, was of great help in recording the phonocardiograms and echocardiograms and Ms Karen Hamilton helped in the preparation of the illustrations and bibliography. Ms Beverley Collins typed the several drafts of the manuscript and her help is gratefully acknowledged. I should also like to express appreciation to Ms Yen Ho, research assistant at the Brompton Cardiothoracic Institute, who prepared the diagrams.

Pittsburgh, 1981 J.R.Z.

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Introduction

Congenital heart disease was once regarded as a medical curiosity; rare, difficult to diagnose during life and impossible to treat. Over the past four decades a very different picture has emerged. Congenital heart disease is not rare, can be precisely diagnosed during life and, in many cases, can be effectively treated. It is, of course, less common in the population as a whole than arteriosclerotic or hypertensive cardiovascular disease, but is now more common than newly diagnosed rheumatic heart disease, at least in more highly developed countries. Congenital cardiac anomalies are estimated to occur in about 8 of every 1000 live births.

The importance of congenital heart disease is enhanced by its high rate of morbidity and mortality, most particularly in early infancy. It has been estimated that 50 percent of newborns with congenital cardiac anomalies will die in the first year of life if left untreated, and even with treatment congenital heart disease is an important cause of death in infancy. Congenital heart disease is, in fact, the single most common cause of death during the first week of life at Children's Hospital of Pittsburgh and accounts for one third of the deaths in this age group. The newborn with congenital heart disease has a more tenuous hold on life than the octogenarian with symptomatic coronary artery disease.

Diagnostic and therapeutic methods have evolved in parallel. The development of surgical techniques for the palliation or repair of cardiac anomalies stimulated the development of diagnostic techniques, and as precise diagnosis became possible during life surgeons were encouraged to extend their horizons and repair anomalies once considered uncorrectable. Although the physiologic consequences of cardiac anomalies are best defined in the cardiac catheterization laboratory, and the detailed premortem elucidation of the abnormal anatomy is by angiography and echocardiography, these techniques have not eliminated the need for a careful physical examination. Indeed, they have enhanced its value by clarifying the genesis of the abnormal physical findings associated with cardiac anomalies. Examples include the echocardiographic demonstration of the origin of the midsystolic click and late systolic murmur of Barlow's syndrome, the 'sail' sound of Ebstein's anomaly and the Austin Flint murmur of severe aortic regurgitation. Another instance is the elucidation of the determinants of width of splitting of the second heart sound by the use of micromanometer tipped catheters in the catheterization laboratory. The implications of physical findings are now more clear and the physical examination has the potential of providing more useful information than it did before the advent of these diagnostic techniques. There is, perhaps, less stimulus to do a thorough physical examination, in the expectation that cardiac catheterization will 'reveal all'. But this is unfortunate, since clinical and invasive methods compliment each other. A physician does a perfunctory physical examination on a child with congenital heart disease at his patient's peril.

Undeniably, the physical examination plays a preeminent role in the recognition of congenital heart disease, since the diagnosis is usually first suspected when a murmur is heard or when cyanosis or congestive heart failure is noted.

It is important to recognize at the outset that congenital heart disease cannot be divided into a number of discrete anomalies or combinations of anomalies, each with specific clinical findings.

Rather, there are spectra of variations of anatomy and physiologic state with corresponding spectra of clinical findings. For instance, there is no such thing as a 'typical' ventricular septal defect with a 'typical' systolic murmur. There may be a more or less 'typical' murmur of a small defect in the area of the membranous septum, or of a tiny defect in the muscular septum, or of a large defect with elevated pulmonary artery pressure, but not of a ventricular septal defect in the general sense. Similarly, there is no 'typical' clinical picture of tetralogy of Fallot. It too is a spectrum, ranging from the nearly asymptomatic young adult with no cyanosis to the desperately ill and severely cyanotic infant. There is a corresponding spectrum of auscultatory findings and both spectra ultimately depend upon the degree of severity of right ventricular outflow tract obstruction. Anatomic variation, pathophysiology, physical findings and clinical course are inextricably intertwined, and heart disease in the child can be best understood when all are considered together.

The physical findings expected with an anomaly can be profoundly altered by the presence of an associated defect. With increasing severity of isolated pulmonic stenosis, for instance, one expects the systolic murmur to become louder and longer, sometimes extending well beyond the aortic closure sound. If there is an associated ventricular septal defect (tetralogy of Fallot) the murmur instead becomes progressively softer and shorter with increasing obstruction, as more and more of the right ventricular stroke volume is shunted through the ventricular septal defect into the aorta rather than traversing the right ventricular outflow tract. In tetralogy of Fallot the murmur may actually disappear during a severe hypoxemic spell, when the right ventricular infundibulum is virtually sealed.

Not all children with congenital heart disease need undergo cardiac catheterization. Although selection of patients for invasive study may depend upon symptoms or electrocardiographic or chest roentgenographic abnormalities, the physical examination is equally important in the selection process. It is quite unusual, for example, to find cardiac symptoms in a child who does not have some indication of severe cardiac disease on physical examination. Further, an asymptomatic child with a normal chest roentgenogram and normal electrocardiogram may have congenital heart disease serious enough to require surgical repair, and in such a child abnormal physical findings form the only basis for selection for cardiac catheterization. As an example, slowly rising arterial pulses, a loud systolic ejection murmur and thrill at the high right sternal border, and an apical early systolic ejection sound indicate severe valvar aortic stenosis. Complete absence of femoral pulses and a right arm blood pressure of 180/100 constitute a severe coarctation of the aorta, even in the absence of symptoms, electrocardiographic abnormality or cardiomegaly. In combination, a soft systolic ejection murmur at the high left sternal border, fixed splitting of the second heart sound and a short mid diastolic murmur at the low left sternal border identify an interatrial septal defect which is large enough to warrant repair, even in a child who seems quite healthy. Accurate clinical diagnosis and a knowledge of the physical findings expected at the more severe end of the spectrum of an anomaly are vital to proper selection for invasive study.

Even when cardiac catheterization has been decided upon, it is quite impossible to plan to use every available hemodynamic and angiographic technique in a particular patient. Selection of the studies most likely to provide pertinent and necessary information is possible only if the physician performing the catheterization has also made a careful clinical evaluation of the patient. The study is likely to be safer and shorter if planned and executed in the light of the clinical findings. In addition, clinical findings serve as a check on data collected in the catheterization laboratory. Even in the best laboratories fallacious data are occasionally gathered, and even the most experienced cardiologist occasionally misinterprets hemodynamic data or angiograms. For example, the extent of pulmonary vascular disease may be underestimated by reliance on 'numbers' alone. The patient with a ventricular septal defect and pulmonary hypertension who has a pulmonary to systemic flow ratio of 1.5 while breathing room air and 2.5 while breathing oxygen may seem to be an acceptable, although not ideal candidate for surgical closure of the defect. If that same patient is known to have a loud single second heart sound at the high left

sternal border, a short soft systolic murmur, a hemoglobin of 17g percent and a history of cyanosis with exertion, enthusiasm for surgical intervention should quickly fade. It must be realized that if the catheterization laboratory diagnosis is not compatible with the clinical picture, both the catheterization data and the physical findings should be carefully reviewed. Conclusions based on a physical examination done with care and system should not be disregarded lightly.

In a book of this sort it is difficult to illustrate adequately with the written word physical findings which depend on hearing, palpation or visible motion. An hepatic pulse tracing does not immediately translate to the 'feel' of a pulsating liver, nor does a giant 'A' wave in the jugular venous pulse tracing give quite the same impression as seeing the real thing. Nonetheless, it is as close as one can come. It is impossible to get any of the 'quality' of a murmur into a phonocardiographic illustration but the shape of a murmur can be shown and the hemodynamic events which produce the murmur, or the abnormally split second heart sound, or the gallop can be illustrated. There remains, however, a large gap between the intellectual understanding of how a particular physical finding came to be and actually appreciating that finding in a patient. The gap can be bridged only by clinical experience.

2

History

The most important function of history taking in a child with heart disease is an assessment of the impact of the cardiac anomaly on the child. The most obvious effects include physical disability, poor growth and development, actual congestive heart failure, limited exercise tolerance, and cyanosis. Less well recognized but sometimes equally important is the emotional impact, ranging from frustration at inability to participate in competitive sports to serious depression or alienation. Other important functions of the history include noting the age of the child when the cardiac anomaly was recognized, searching for a family history of congenital heart disease and assessing compliance with medical instructions.

History taking must be directed, to a degree, since certain areas must routinely be explored and may be missed if the parents are not asked specific questions. Almost all parents are eager to give as much information as possible about their child, but the physician may be tempted to turn off or redirect the conversation when it strays from a response to a direct question. Somewhere in the course of history taking, however, there should always be a time when parents are allowed, indeed encouraged, to tell what seems to them to be important about their child. Not only may valuable information about disability emerge, but one will also get a better idea of the parents' perception of the child's state, opening the door to more effective counselling and education. It is important for parents to get as clear an idea as possible of the nature of their child's anomaly and its seriousness and possible consequences. The physician can accomplish this best if he understands the parents' current knowledge, anxieties, perceptions and misconceptions about their child's problem.

Although infants with congenital heart disease may grow slowly, the poor growth may or may not be causally related to the cardiac anomaly. Indeed, unless there is a large left to right shunt with cardiomegaly or unless there is arterial unsaturation, slow growth is usually not *caused* by cardiac disease. (To put it another way, there will not be a growth spurt following repair of the defect.) Even if there is cardiomegaly and/or cyanosis, growth may not improve after successful surgery. The symmetry, or lack of it, of the growth pattern is a useful differential point, since weight, height and head circumference are not equally affected by severe congenital heart disease. Of these variables, weight is most likely to be influenced by cardiovascular stress, and if weight is below the 5th percentile while head circumference is normal the cardiac anomaly is very likely responsible for the poor growth. On the other hand, if head circumference, weight and height are all below the 5th percentile none are likely to be attributable to cardiac disease.

Parents of a child with suspected congenital heart disease should always be asked how their child breathes, how he eats, how he has gained weight and whether or not he has ever been blue, since important evidence of heart failure or of arterial oxygen desaturation may be obtained during the taking of the history. The infant in congestive heart failure feeds poorly; he typically takes a small amount of milk, rests or falls briefly asleep, and then wakes hungry and begins feeding again. With severe congestive heart failure, the infant may stop feeding entirely.

The infant with congestive heart failure may be tachypneic at rest or even after having fallen asleep. It is unwise to dismiss a parental report of rapid

breathing without making an especially careful search for other evidence of heart disease, especially if the history is given by an experienced parent or by a grandmother. Unfortunately, it is not uncommon to hear from a parent that an infant now in obvious congestive heart failure had been tachypneic for some time before overt failure appeared and that the breathing pattern had been dismissed by nursery personnel or by a physician as being of no consequence.

Parents may not be aware of cyanosis in their child even if it is quite obvious to the examiner. This is especially likely if the cyanosis is constant and has developed gradually. Parents are likely to report episodic cyanosis, however, especially if it is accompanied by the respiratory distress or change in level of consciousness which occur in the hypercyanotic spells of tetralogy of Fallot (or less commonly with other varieties of cyanotic congenital heart disease). It is important to note that not all episodic cyanosis is cardiac; normal infants as well as older children may display obvious cyanosis of lips or nail beds when exposed to cold. (Parents frequently report that perfectly normal children become cyanotic after swimming.)

It is important to inquire as to the adequacy of iron intake in infants who are or who are expected to be cyanotic. If iron intake is inadequate the resulting anemia (which may be absolute or relative) may increase symptoms by reducing the oxygen carrying capacity of the blood. With tetralogy of Fallot, for instance, symptoms may be very mild with a hemoglobin level of 18 g percent, but become severe and include hypercyanotic spells if the hemoglobin drops to 10 g percent. Anemia, especially the microcytic variety which occurs with iron deficiency, may precipitate a cerebral vascular accident in a child with cyanotic congenital heart disease.

Symptoms may or may not be present in the older child with highly significant congenital heart disease. With purely obstructive lesions such as pulmonic or aortic stenosis or isolated coarctation of the aorta, symptoms tend to occur very late. In addition, children have a remarkable capacity to adapt and may not be aware of limitations when they indeed exist. The older child may also display a strong tendency to deny limitations and may compensate by attempting to maintain a high level of physical activity. On the other hand, over concerned parents frequently report easy fatigability and exercise intolerance in children with anomalies which are hemodynamically unimportant. Almost invariably the child with cardiac symptoms has other evidence of the severity of his anomaly, either on physical examination, chest roentgenogram, or electrocardiogram. (Important exceptions to this general rule include paroxysmal dysrhythmia, hypertrophic cardiomyopathy and intracardiac tumor.) If cardiac symptomatology seems inconsistent with the remainder of the clinical examination, observation of the child during exercise, either casual or during a formal exercise study, may be quite helpful. It is worth noting that chest pain in children is very rarely cardiac in origin. This fact should be made known to the worried parent.

Although episodes of syncope or of seizure activity in children are not usually of cardiac origin, the possibility of an episodic arrhythmia or of the 'long QT' syndrome should be considered in children with otherwise unexplained 'spells' and should lead to a careful cardiac physical examination and an electrocardiogram.

Although the history is less important in establishing the presence of organic heart disease in a child than it is in the adult with coronary artery disease, it is an invaluable source of information, particularly with regard to the impact of the cardiac anomaly on the child's physical state and psyche.

3

The cardiac physical examination

Three indispensable prerequisites to a good physical examination are a proper environment, a cooperative child and a conviction on the part of the physician that the examination is important. An ideal environment includes quiet and the opportunity for both the child and the physician to be comfortable. Both are easily obtained in an office setting, but may be more difficult in the hospital, where noise levels are often high and rooms crowded. Radios, television sets and conversing colleagues are common sources of ambient noise, and each should be 'turned off' before the examination begins. Most physicians find that a physical examination is most easily performed from the patient's right side, and it is better to move a bed or chair than to alter one's customary approach. A patient should always be examined from a comfortable position if at all possible.

Obtaining the cooperation, or at least the tolerance of the child may require considerable patience and skill on the part of the physician. There is usually little problem with the newborn or small infant, who is usually asleep or at least docile if well fed and dry. The older infant and young child may pose more of a challenge, and it is well to begin by asking the mother whether the child is more likely to be happy in her lap than on an examining table. It is possible to do a thorough cardiovascular examination with an infant sitting and lying in his mother's lap, and better to begin that way if the child is likely to object to the separation of an examining table. If the table is tried first and the child vigorously resists, the chances of success with a subsequent attempt in the mother's lap are appreciably lessened.

It is worthwhile getting acquainted with the child before any 'laying on of hands'. This may require little more than a smile with a six-month-old but may be more involved with an older child. Sitting down beside the mother and child and giving the child a small toy or even a tongue blade to play with is less threatening than a direct approach with a stethoscope, a strange and sometimes frightening object to a two-year-old. If the child seems to fear the stethoscope, he may be persuaded to hold and examine it by his mother and may then submit to being examined. Also, auscultation may be less threatening if a toe, leg or the abdomen is 'listened to' before the chest. (The stethoscope head should be warmed with the hand before touching the child with it.) The importance of a nonthreatening approach cannot be overestimated; a smiling and unhurried examiner is much more acceptable to a toddler than a solemn or abrupt one.

Occasionally, even with a gentle approach, young children remain uncooperative. The infant who cries vigorously during an attemped examination may respond to a bottle. Failing this, he may fall asleep if the examiner leaves the room and may remain asleep during auscultation after the examiner returns. If a window is available, a refractory toddler can sometimes be distracted by drawing his attention to a bird or cat, real or imaginary, outside the window. It is surprising how long the charade can be continued and how much information can be obtained using this simple artifice. A pre-schooler may be persuaded to cooperate simply by telling him he is 'expected' to be quiet. In the author's experience it has rarely been necessary to sedate a child to perform an adequate physical examination.

The physical examination is easily the most important non-invasive diagnostic tool for the

detection and evaluation of heart disease in children. The physician who does a perfunctory physical examination, either because of lack of skill or because of his hurry to proceed to 'more objective methods' such as the chest roentgenogram, electrocardiogram or echocardiogram fails to make full use of readily available and potentially valuable information. In this chapter, the parts of the cardiac physical examination will be outlined and the physiologic basis for the various findings will be explored. When appropriate, reference will be made to other portions of the text for specific details.

THE JUGULAR VENOUS PULSE

The normal jugular venous pulse consists of 'A', 'C' and 'V' waves. The 'A' wave is caused by right atrial contraction, the pressure wave being transmitted through the venous system to the jugular veins. The 'C' wave occurs at the beginning of ventricular systole and usually appears as a hump on the descending limb of the 'A' wave. It is probably a combination of an impulse transmitted from the nearby carotid artery and a pressure wave generated by bulging of the closed tricuspid valve leaflets into the right atrium during very early ventricular systole. The 'X' descent follows the 'C' wave. It is largely due to the tricuspid valve annulus being pulled away from the right atrial cavity by the contracting right ventricle, reducing right atrial pressure in the process. As venous return continues through systole there is a slow increase in right atrial pressure, visible in the jugular veins as the 'V' wave. The 'V' wave peaks just as the tricuspid valve opens. As right atrial blood enters the right ventricle through the opened tricuspid valve, the 'Y' descent begins.

In adults it is commonly possible to see and record all of the above waves. In infants and children the venous pulse is much harder to see, both because of the shorter neck and more abundant subcutaneous tissue of the young child and because the faster heart rate common in younger individuals blurs the distinction between the waves and makes them harder to interpret. It is thus rarely possible to identify all of the 'normal' jugular venous waves in an infant. In some children and in an occasional infant an abnormal jugular venous pulse can be seen if looked for and may then give valuable insight into right heart hemodynamics.

Venous pulsations are usually more marked in the internal than in the external jugular veins, since venous valves in the external system interfere with retrograde transmission of waves from the right atrium. It is important that the neck be relaxed, since tensing of muscles may obscure venous waves. Jugular venous pulsations are strikingly dependent on the position of the patient, specifically on angulation of the trunk from the horizontal, since this angulation determines elevation of the observed vein above the right atrium. The pulsations are most obvious at the top of the 'fluid level' in the vein, or in other words at the level where the vein becomes distended by returning blood. (The vein below this point also must pulsate to a degree, but pulsations are much less obvious.) The proper angle at which the jugular venous pulse is most obvious varies from patient to patient and also from time to time in the same patient, depending upon right atrial pressure levels. Usually the veins are best seen with the patient reclining at a 15 to 30° angle, but when right atrial pressures are very high the pulsations may be seen only with the patient sitting upright. Lighting is very important, and venous pulsations are best seen when light strikes the neck tangentially, throwing the veins into relief. The veins are much less well seen in diffuse light, and if proper window lighting is not available a flashlight can be used to highlight the veins.

Visible pulsations in the neck may be of arterial or venous origin, and the distinction may occasionally be difficult, particularly if venous pulsations are unusually strong. The failure of arterial pulsations to change with position is a useful distinction and, also, venous pulsations can be damped by light pressure above the clavicle, most easily applied by stretching the skin over the area. (If pressure is too heavy, the vein will distend, since return to the heart is blocked. Even so, pulsation is eliminated.) Jugular venous waves can be timed and identified either by palpating the opposite carotid or by simultaneous auscultation of the heart. 'A' waves are, of course, presystolic and precede the carotid pulse and the first heart

sound. 'V' waves are nearly coincident with or closely follow the carotid pulse and occur after the first heart sound.

Prominent 'A' waves result from forceful right atrial contractions and signify increased impedance to right atrial emptying. Such increased impedance is most commonly a result of the decreased right ventricular compliance which occurs with right ventricular hypertrophy (e.g. with severe 'isolated' pulmonic stenosis, primary pulmonary hypertension, or cardiomyopathy). Increased impedance to right atrial emptying also occurs with tricuspid stenosis or atresia and, in the latter entity, large jugular venous 'A' waves are presumptive evidence of a restrictive interatrial communication (Fig. 3.1). Still another cause of increased impedance to right atrial emptying is the concurrence of atrial and ventricular systole, atrial contraction then occurring while the tricuspid valve is closed. The resultant high right atrial pressure is transmitted to the jugular veins and is known as a 'cannon' wave (Fig. 3.2), one of the characteristic findings

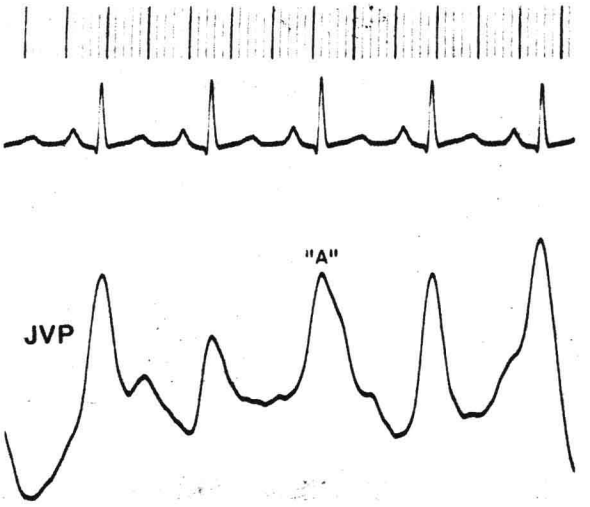
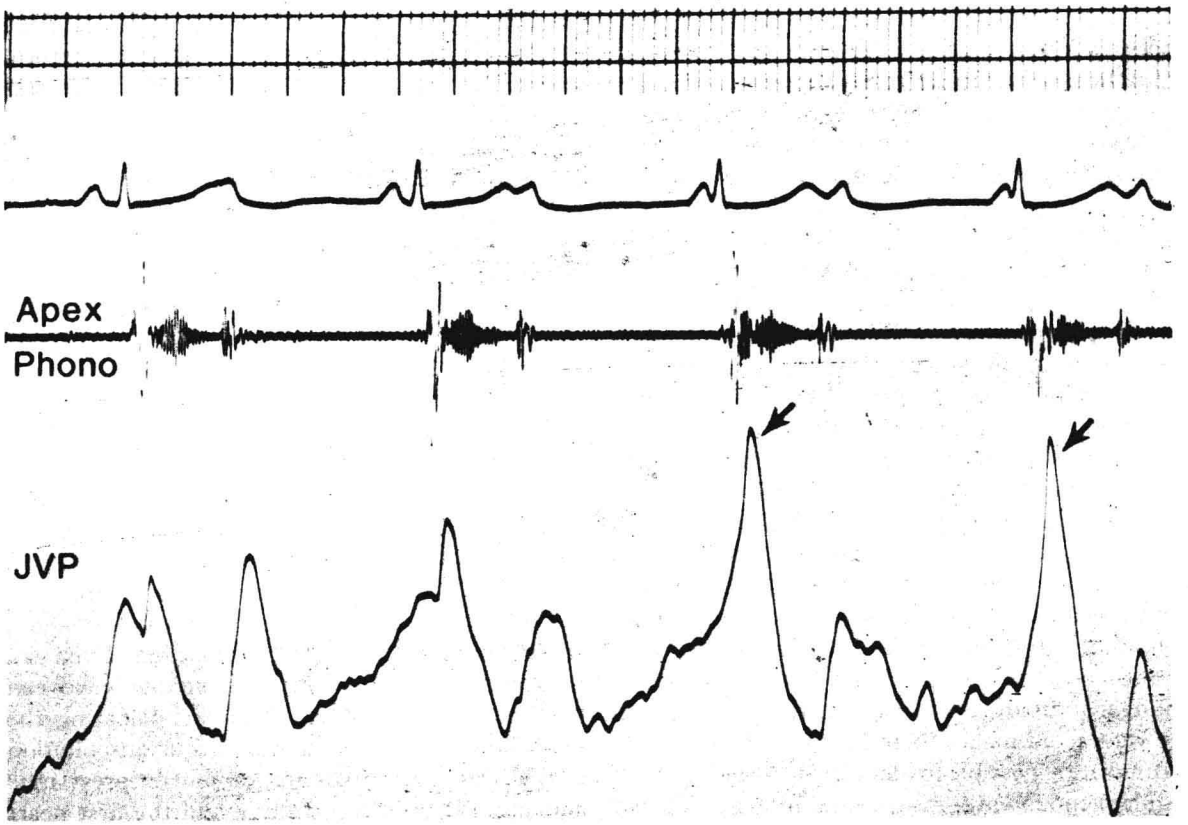


Fig. 3.1 Child with tricuspid atresia and restrictive interatrial opening. Large 'A' waves are evident in the jugular venous pulse recording (JVP).

Fig. 3.2 Child with congenital third degree atrioventricular block. Cannon waves (arrows) are seen in the jugular venous pulse (JVP) when mechanical atrial systole occurs during ventricular systole (note very short apparent PR interval).



in third degree atrioventricular block. Since atria and ventricles are not electrically linked in this situation, atrial and ventricular contractions will, by chance, occasionally coincide. The intermittent nature of the large waves is characteristic, but can also occur with second degree atrioventricular block or with supraventricular premature beats, the common denominator being coincidence of atrial and ventricular systole. A long PR interval tends to make an 'A' wave more obvious, since it separates it from the subsequent carotid pulse. A short PR interval can actually be the cause of cannon waves, as they were in the early postoperative period in one of our patients who had undergone a Mustard procedure for transposition of the great arteries. Striking neck vein pulsations led to a concern that there might be partial obstruction to superior vena cava return. The presence of a junctional rhythm suggested that the pulsations might, instead, be cannon waves and the latter etiology was established when the large 'A' waves suddenly disappeared with reversion to a regular sinus rhythm.

If atrial and ventricular systole are not concurrent, a large jugular venous pulse coincident with or following the carotid pulse (or after the first heart sound if timing is by auscultation) is a 'V' wave and is generated by right ventricular systole (Fig. 3.3). Pathologic 'V' waves indicate important tricuspid regurgitation. It is important to note that significant tricuspid regurgitation can exist without producing large 'V' waves, since the right atrium enlarges with chronic regurgitation and can absorb considerable regurgitant volume without much change in pressure. The damped right ventricular impulse will not then be evident in the jugular venous pulse.

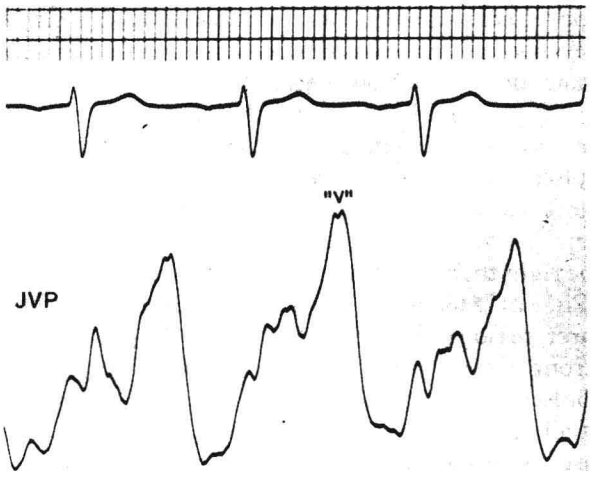


Fig. 3.3 Child with tricuspid regurgitation. A large 'V' wave is present in the jugular venous pulse recording (JVP).

If right atrial 'A' or 'V' waves are sufficiently large the jugular pulsations may be palpated as well as seen. In the author's experience this finding has been most common in patients with either tricuspid atresia and a restrictive interatrial septal defect or with a cardiomyopathy. Palpable venous waves may also occur with severe tricuspid regurgitation and with severe 'isolated' pulmonic stenosis. Large atrial waves may be transmitted to the hepatic veins and may cause the liver to pulsate (Fig. 3.4). Hepatic pulsation must be differentiated

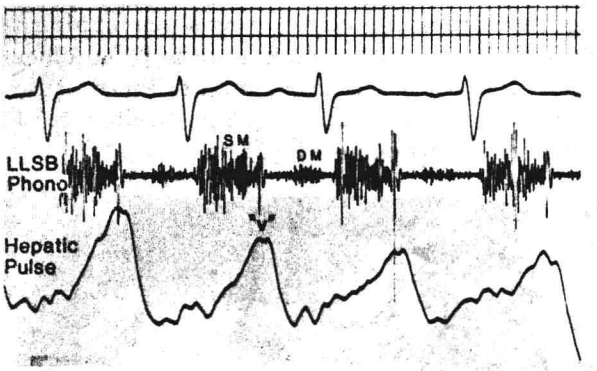


Fig. 3.4 Same child as Figure 2.3. The low left sternal border phonocardiogram shows both the systolic murmur of tricuspid regurgitation and the mid diastolic murmur of relative tricuspid stenosis. The hepatic pulse tracing shows a prominent 'V' wave.

from a transmitted cardiac impulse, and a pulsating ventral surface of the liver, felt just below the right costal margin, is more reliable evidence of large 'A' or 'V' waves than is movement of the lower edge of the liver synchronous with the cardiac impulse.

In adults, abnormal distension of the jugular veins occurs most commonly with congestive heart failure and reflects elevated right atrial pressure. The normal inspiratory fall in central venous pressure, and hence of the level of distension of jugular veins, may be lost in such a patient. If failure is severe there may be an inspiratory increase rather than a decrease in the level of distension (Kussmaul's sign). (It should be noted

that constrictive pericarditis can also produce an inspiratory increase in jugular venous distension.) In patients with borderline right ventricular function, distension may also increase when the right upper quadrant of the abdomen is compressed with the hand, quiet respiration being maintained to prevent a Valsalva maneuver. This hepatojugular reflux occurs in patients whose 'failing' right ventricle is unable to increase its output as the abdominal compression increases systemic venous return.

In infants, congestive heart failure rarely results in obvious jugular venous pulsations or distension. If distended neck veins are present in an infant in failure, the possibility of an intracranial arteriovenous fistula should be considered. In this entity distension is related to a combination of increased right atrial pressure and to torrential venous return through the jugular venous system. In an older child, a cervical arteriovenous fistula can cause jugular venous distention, even in the absence of congestive heart failure (Fig. 3.5).

Fig. 3.5 Distended neck vein (arrows) in a child with a cervical arteriovenous fistula.

ARTERIAL PULSE

Peripheral arterial pulses should be described in terms of amplitude and symmetry. Judging the general amplitude of peripheral pulses is a subjective exercise, but attention to more distal arteries may be helpful in differentiating normal from increased pulses. Strong palmar or digital pulses, for instance, constitute good evidence that pulse pressure is increased. Pulses of increased amplitude can result from low systemic arterial resistance, as with an aortico-pulmonary artery communication (e.g. patent ductus arteriosus, aortico-pulmonary window, or persistent truncus arteriosus) or with a systemic arteriovenous fistula. Pulses are also increased when myocardial inotropy is high and most of the stroke volume is ejected in early systole (e.g. hypertrophic cardiomyopathy). Strong pulses also result from the physiologic increase in cardiac output and decrease in systemic vascular resistance which accompany fever or physical exertion. Peripheral pulses are weak when the central aortic pulse is damped by severe aortic stenosis or when there is congestive heart failure, with its low cardiac output, small stroke volume, and reflexly increased systemic vascular resistance. Striking respiratory variation in pulse amplitude suggests either pericardial tamponade or airway obstruction (Ch. 21).

To judge symmetry of arterial pulses, either the brachial, radial or axillary pulse should be felt and compared in each arm and then compared with the femoral pulses. If leg pulses are decreased or absent, a coarctation of the aorta may be assumed to be present. (Since femoral pulses may be hard to feel in the chubby infant or obese older child, it is well to check dorsalis pedis and posterior tibial pulses as well as femorals.) If a coarctation of the aorta is suspected, a femoral pulse and the brachial or axillary pulse in the right arm should be felt simultaneously. In the normal individual there is rarely enough delay in the femoral pulse to be appreciated at the bedside; with coarctation it may be obvious that the femoral pulse is not only weaker than the brachial but delayed as well (Fig. 9.2). Since the origin of the left subclavian artery may or may not be involved in a coarctation, left arm pulses may be as decreased as the femorals or may more closely resemble pulses in the right arm. Another less common cause of a stronger pulse in the right than left arm is supravalvar aortic stenosis (Ch. 12). Stenosis in a subclavian artery is a still rarer reason for a weak arm pulse. If pulses are strong in the legs but weak in the arms Takayasu's (pulseless) disease is suggested. If pulses are weak in all four extremities several possibilities should be considered. The most likely cause is low cardiac output, either because of poor myocardial function or because of obstruction to left ventricular outflow

(e.g. aortic atresia or very severe aortic stenosis). A rare but curable cause of decreased pulses in both arms and both legs is a coarctation of the aorta which involves the left subclavian artery and which is associated with distal origin of the right subclavian artery. In this situation carotid and temporal pulses are strong and make the diagnosis.

BLOOD PRESSURE

Blood pressure determination is an important part of the physical examination of the child, particularly since some causes of childhood hypertension are curable and since the first evidence of adult essential hypertension may appear during childhood or adolescence.

Accurate blood pressure determination demands attention to a few details. The cuff width should be adequate, covering at least two-thirds of the upper arm. A small cuff leads to a spuriously high blood pressure estimate and should be avoided. A cuff which covers the entire upper arm from the axilla to the elbow does not distort blood pressure and is quite acceptable. The cuff should be inflated until distal arterial pulses disappear, and then deflated until Korotkoff sounds appear over the brachial artery just below the cuff, marking systolic blood pressure. With further deflation the sounds either disappear or abruptly fade, marking diastolic blood pressure. In infants, the flush technique has been advocated and does give a fairly good measure of blood pressure. In this technique, blood is expelled from the hand and forearm as the cuff on the upper arm is inflated. As cuff pressure is gradually lowered, arterial blood suddenly enters the lower arm and the skin becomes pink and 'flushed'. Even in small infants, however, blood pressure can usually be determined by auscultation or palpation and is almost certainly more accurate than blood pressure determined by the flush technique.

Ideally, blood pressure should be determined in both arms and in a leg. This is mandatory if coarctation of the aorta is suspected because of pulse asymmetry or hypertension. If a coarctation is present, blood pressure is lower in the legs than in the right arm, while pressure in the left arm is variable, depending upon whether or not the origin of the left subclavian artery is involved in the coarctation. In children, hypertension may also be due to renal arterial narrowing or renal parenchymal disease, or be of undetermined etiology (essential hypertension). Asymmetry of blood pressure is most commonly related to coarctation of the aorta, but may also accompany supravalvar aortic stenosis (Ch. 12). In this entity pressure in the right arm may be higher than in the left. (Even in normal individuals, right arm pressure may be higher than left arm; the asymmetry of supravalvar aortic stenosis is simply an exaggeration of this normal variation. To be significant, the difference in right and left arm systolic pressure should exceed 10 mmHg.) Low blood pressure is infrequently a clinical problem, unless an infant or child is obviously extremely ill and has a very low cardiac output.

Blood pressure normally varies slightly with respiration, rising with the increased intrathoracic pressure of expiration and falling as intrathoracic pressure falls with inspiration. Respiratory variation is recognized by inflating the blood pressure cuff until Korotkoff sounds disappear, then slowly deflating the cuff until sounds appear during the expiratory phase of quiet respiration and mark the upper limit of systolic blood pressure. The cuff is further deflated until Korotkoff sounds are present through the respiratory cycle, marking the lower limit of systolic blood pressure. The difference between the upper and lower limits is the measure of respiratory variation and normally does not exceed 10 mmHg. A respiratory variation in excess of 20 mmHg (paradoxical pulse) strongly suggests either pericardial tamponade or airway obstruction (Ch. 21).

PRECORDIAL MOTION

The beating heart moves within the thorax, and in some incompletely defined manner imparts movement to the thoracic wall. In the normal individual the chest wall motion is small and is well localized. The major movement (point of maximal impulse—PMI) overlies the cardiac apex, usually in the fourth or fifth intercostal space near the mid clavicular line, and is related to left ventricular contraction. Normally, there is a brief systolic outward movement which peaks at the beginning