Left Ventricular Hypertrophy Syndrome in Infancy

SIDNEY BLUMENTHAL, M.D., F.C.C.P. and SAMUEL O. SAPIN, M.D.

New York, New York

Cardiac hypertrophy in infancy has been the subject of considerable interest for many years. An increasing number of reports have appeared in the literature describing diverse cardiac lesions in infancy, which nevertheless have similar clinical manifestations. Rosenbaum et al. incorporated this group of diseases under the heading "Primary Myocardial Disease in Infancy and Childhood." Since most of these lesions are not of primary myocardial origin and since they produce left ventricular hypertrophy as the predominant anatomic finding, we offer "Left Ventricular Hypertrophy Syndrome" as a more descriptive title.

The Left Ventricular Hypertrophy Syndrome in infancy is characterized by: (1) cardiac enlargement—predominantly of the left ventricle; (2) absence of significant heart murmurs; (3) absence of central cyanosis; (4) electrocardiographic findings which are either normal or indicative of myocardial damage or of left ventricular hypertrophy; (5) at times, symptoms and signs of cardiac failure.

The lesions associated with this syndrome are conveniently divided into those in which the predominant abnormality is endocardial, as in primary endocardial fibroelastosis; those in which the myocardium is primarily involved and a third group in which there is predominant alteration in function of the coronary arteries.

A number of rather common congenital malformations producing left ventricular hypertrophy in the absence of cyanosis are seen in infancy. These are usually accompanied by significant murmurs and, by definition, are excluded from the syndrome under discussion. They include patent ductus arteriosus, ventricular septal defect, aortic or subaortic stenosis as well as coarctation of the aorta.

In a characteristic instance of the left ventricular hypertrophy syndrome, the history is that of a previously healthy baby who has begun to eat poorly, and to show signs of restlessness and fatigue. Episodes of apparent pain may be noted. Restlessness increases and is often followed by respiratory distress. On examination, dyspnea, tachycardia, enlargement of the heart and, in some instances, gallop rhythm and poor heart sounds are noted. Hepatomegaly is often present due to associated cardiac failure. Cyanosis, if present, is usually transient or terminal. Heart murmurs are conspicuously absent or, if present, consist of soft, inconsequential, apical murmurs. Usually the lungs are clear but evidence of pulmonary infection or signs of atelectasis produced by cardiac enlargement may be present. Peripheral edema is uncommon except as a late manifestation. Radiologic examination reveals considerable cardiac enlargement, the silhouette often being globular in shape; occasionally the standard criteria for left ventricular enlargement are satisfied. The electrocardiogram may be diagnostic of left ventricular hypertrophy or reveal ST or T wave abnormalities indicative of myocardial anoxia or damage.

From the Department of Pediatrics, College of Physicians and Surgeons, Columbia University and the Department of Pediatrics, Mount Sinai Hospital.
We shall discuss the lesions resulting in the left ventricular hypertrophy syndrome as defined, describing their characteristics, differential diagnosis and treatment.

I. Primary Endocardial Disease

a. Primary Endocardial Fibroelastosis is the most common cause of the left ventricular hypertrophy syndrome in infancy. Kelly and Anderson\textsuperscript{2a} collected 79 cases from the literature and added 17 from the files of the Babies Hospital. It is apparent that a larger number of cases have never been reported. Many were formerly classified as idiopathic cardiac hypertrophy of infancy, while in others the cause of death may have been overlooked completely. Kugel and Stoloff\textsuperscript{3} were among the first to recognize the significance of endocardial thickening in patients described as having idiopathic cardiac hypertrophy. Andersen and Kelly\textsuperscript{2b} emphasized the importance of separating cases of endocardial fibroelastosis into two groups: those without associated cardiac malformations (primary endocardial fibroelastosis) and those associated with congenital malformations in which the endocardial thickening is secondary to hemodynamic changes related to the cardiac malformation. The latter group have been excluded from this discussion as they are not representative of the left ventricular hypertrophy syndrome.

The cause of primary endocardial fibroelastosis is not well understood. Gross\textsuperscript{4} was one of the first to point out that there is no evidence of an inflammatory process. He felt that simple hyperplasia of the endocardial fibroelastic tissue best explained the condition, and that it was a developmental defect rather than an inflammatory process. Others\textsuperscript{5} have supported this interpretation. The concept that endocardial anoxia may be the stimulus for the production of endocardial fibroelastosis has been suggested by several authors. Thomas et al.\textsuperscript{6} indicate that endocardial fibroelastosis associated with other cardiac malformations is confined to patients in whom a clear-cut cause for endocardial anoxia is demonstrated. Craig\textsuperscript{7} states that the myocardial degeneration and fibrosis so frequently encountered in the subendocardium is the result of such anoxia. Johnson\textsuperscript{8} suggests that, in the primary variety, endocardial anoxia in the left heart may be present in utero due to either delayed opening of the septum primum or early closure of the foramen ovale. The possibility that this is a developmental anomaly has been suggested by the familial incidence in reported cases such as in siblings and multiple pregnancies.\textsuperscript{2, 7, 10, 11} In only an occasional case has a specific prenatal maternal illness been present.\textsuperscript{9}

Assuming that the original cause is either endocardial anoxia or a developmental defect, Weinberg and Himmelfarb\textsuperscript{10} propose that the thickened endocardium may prevent adequate nourishment of the myocardium. Drainage of the arterioluminal vessels into the ventricle is obstructed so that partial stasis occurs in intramyocardial capillaries. This results in myocardial anoxia and subsequent myocardial failure. Kelly and Andersen\textsuperscript{2a} suggest a possible congenital familial metabolic defect resulting in myocardial weakness with endocardial fibrosis as a secondary phenomenon. They indicate that investigations directed towards a deficiency of some enzyme involved in myocardial metabolism may be a profitable approach to the problem.
The clinical findings in primary endocardial fibroelastosis are similar to those described for the entire left ventricular hypertrophy syndrome. Significant heart murmurs are absent in about 80% of all cases. Cardiac hypertrophy is demonstrable by physical and roentgen examinations. The electrocardiogram reveals no pathognomonic features which distinguish this entity from the other lesions of the left ventricular hypertrophy syndrome. Standard leads in the electrocardiogram often reveal evidence of nonspecific myocardial anoxia or damage with T wave flattening or inversion or, less often, with ST segment deviation. Many of the precordial tracings available in the literature indicate the presence of left ventricular hypertrophy. Deep Q waves with marked ST segment deviation are unusual in fibroelastosis. In some instances conduction defects or paroxysmal tachycardia have been noted.

Cardiac catheterization has been performed in a few patients with proven endocardial fibroelastosis. The right ventricle and pulmonary artery pressures have been normal or slightly elevated with elevation of the pulmonary capillary pressure. The features of constrictive pericarditis have been reported in adults with severe endocardial fibrosis. This has not been a finding in fibroelastosis in infancy. Angiocardiograms reveal poor emptying of the left ventricle. This finding might support either the theory that the heart fails in endocardial fibroelastosis because the thick endocardial coating prevents proper cardiac contraction or that of primary muscle dysfunction.

Primary endocardial fibroelastosis is essentially a disease of infancy. Some cases survive into childhood, and an eleven-year-old child has been reported by Blumberg and by Thomas. A pathological counterpart of endocardial fibroelastosis similar to the primary syndrome of infancy has been noted in adults and reported under various titles. However, it is not clear whether these cases indicate survival following the infantile syndrome or represent a separate entity. The course of the disease in infants may be somewhat variable. Eighty percent of the patients die by the age of one year. We have observed two patients who succumbed during the neonatal period. Some infants die suddenly. Others improve temporarily with digitalis, oxygen and supportive therapy only to relapse on one or several occasions. In rare instances the improvement may continue and the patient reach childhood at which age the diagnosis of endocardial fibroelastosis can only be suspected. One is not justified in excluding this diagnosis in a given case merely on the basis of survival.

At post mortem examination one notes thickening of the mural endocardium involving both the fibrous and elastic tissues. The endocardium of the left ventricle is most frequently involved, that of the left atrium less so. Occasionally the process extends to the right side of the heart, involving both left and right, and in a rare case the endocardium of the right heart alone has been thickened. The process may extend to involve the valves which then become thickened and deformed. When valvular involvement exists, the mitral and aortic leaflets are most commonly affected. However, almost all valvular combinations have been described. There is considerable thickening of the myocardium which, in about 80% of the reported cases, is limited to the left ventricle. The myocardial changes include degenerative processes, fibrosis and calcific deposits. The
alterations are most prominent in the sub-endocardial area and in the papillary muscles. Occasionally there may be perivascular hyperelastosis and fibrosis of the coronary arteries and also of the arteries in other organs.5

II. Primary Myocardial Disease

a. Idiopathic Myocarditis: This is a relatively uncommon disease in infancy characterized by an acute inflammatory involvement of the myocardium without any recognizable associated disease. It resembles the myocarditis seen as a complication of the acute infectious diseases. Pathologically there is hypertrophy and dilatation which is generalized or at times confined to the left ventricle. In one of our cases the degree of myocardial involvement was so extensive as to result in the formation of a ventricular aneurysm. Endocardial thickening may be a secondary phenomenon. Microscopic examination reveals evidence of acute inflammation as well as fibrosis and scarring. Aschoff bodies are not demonstrable and the valves are spared. The coronary arteries are normal.

Idiopathic myocarditis is probably caused by a variety of agents including bacteria and viruses. At times its distribution has been epidemic in character. The possibility of a viral etiology has recently been emphasized. Stoeber22 reported 140 instances of this disease in infancy in which diffuse myocarditis was noted with little or no endocardial or pericardial involvement. A viral etiology was suggested but not proved. Amongst these cases the highest incidence was at approximately one year of age. Montgomery23 reported a group of newborns with acute myocarditis with isolation of Coxsackie group B virus from the feces of one fatal case and of one survivor. Javett et al.24 reported an outbreak amongst newborns in South Africa in 1952. Ten infants were diagnosed as having acute myocarditis of whom six died. Coxsackie group B type 3 virus was isolated from the feces of one of the survivors. In a subsequent similar case they were able to recover this virus from the myocardium. A Coxsackie virus has also been isolated from the myocardium by Van Crevel26 and by Kibbrick and Benirschke26 working in Enders' laboratory. The latter investigators isolated Coxsackie group B type 3 virus from the spinal cord of an infant dying from myocarditis on the seventh day of life and type 4 from the myocardium of a 10 day old infant. They postulate, as did Lind,27 that the disease might be acquired in utero by way of transplacental transmission from a mother with a minor respiratory illness. Enders indicates that the Coxsackie B virus is the cause of some cases of acute aseptic myocarditis in infancy in addition to those cases of meningitis or encephalomyelitis in which there is associated myocardial involvement.

The clinical picture is similar to that caused by other conditions resulting in left ventricular hypertrophy in infants. The onset is sudden and the entire course may be a rapidly fatal one. Some are noted to have a relatively short biphasic pattern while, in others, a more chronic course measured in months has been noted. The disease varies markedly in severity and duration. Reports indicate that in the nonfatal cases recovery is complete without valvular or myocardial residua. Fluoroscopy and radiologic examination reveal general cardiac enlargement. The electrocardiogram demonstrates ST segment and T wave changes consistent with
myocardial anoxia or damage. In some, conduction defects are noted. The pattern of massive myocardial infarction has been simulated in one of our cases. The type and severity of the electrocardiographic changes are dependent upon the site and degree of myocardial involvement. Miller\textsuperscript{29} believes that "a dependable electrocardiographic pattern of myocarditis" exists. It would seem that more data are necessary to substantiate this opinion. In our experience the electrocardiographic findings usually are of a non-specific nature similar to those seen in other conditions in the left ventricular hypertrophy syndrome.

b. Glycogen Storage Disease of the Heart is an exceedingly rare disorder resulting in the deposition of large amounts of glycogen in the myocardium and in striated muscle. This is commonly a familial disorder. The patients are normal at birth but acquire symptoms during early infancy. They gain and develop poorly. Tachycardia and dyspnea are early signs and tremendous cardiac enlargement is an early finding. Murmurs are conspicuous by their absence. Di Sant'Agnese et al.\textsuperscript{30} noted macroglossia in several cases. Laboratory tests directed at evaluating glucose tolerance and glucose mobilization are normal. Ketonuria is not a feature of this disease. Roentgen findings are those of non-specific generalized cardiac enlargement. The electrocardiographic changes include T wave abnormalities similar to those found in other diseases of the left ventricular hypertrophy syndrome. In some, electrocardiographic evidence of left ventricular hypertrophy has been noted, but too few precordial lead studies are available to indicate the frequency of this observation. Death may occur suddenly or after an episode of congestive failure, usually before the end of the first year of life.

This disease is distinctly different from the type of glycogen storage disease in which the liver is predominantly involved. There is no gross involvement of the liver, but microscopic and chemical analysis reveal abnormal glycogen deposits in a large number of organs of the body. Abnormal amounts of glycogen are deposited in the myocardium and in striated muscle. The heart has a distinctive post mortem appearance as noted by Andersen. It is enormously enlarged, round in shape, with a homogeneous appearance of the myocardium. Microscopically the myocardial fibers are very large and vacuolated. The pathogenesis of this metabolic disease is poorly understood.

A family history indicating that other infants had died under similar circumstances as well as the finding of cardiomegaly and macroglossia should suggest cardiac glycogen storage disease as a diagnostic possibility. Skeletal muscle biopsy should be performed and may reveal excess glycogen storage. At the present time the prognosis is universally bad. There is no satisfactory treatment for this disease.

c. Tumors: Primary tumors of the heart are rare in infancy. The most common type is the rhabdomyoma. Kidder\textsuperscript{31} found a total of 69 such tumors in his review of the literature; half were discovered in the first year of life. They are often found in patients with tuberous sclerosis. The heart is enlarged and the characteristics of the left ventricular hypertrophy syndrome may be noted. Murmurs may or may not be present depending upon whether the tumors are in or upon a valve with resultant
obstruction of blood flow. Arrhythmias have been noted frequently. As a rule, death occurs as a sudden event.

Fibroma of the myocardium, myxoma of the atrium and rhabdomyosarcoma have also been observed in infancy. A recent report describes a rhabdomyosarcoma in an infant four months of age in whom the myo-cardium and left coronary artery were involved. Electrocardiographic signs of non-progressive, marked ST segment elevation in the precordial leads V2-6 was present in this infant with cardiac failure resistant to medical therapy.

d. Nutritional Deficiencies: It is well known that diets deficient in vitamins may produce myocardial damage in experimental animals and in humans. The human heart is affected in thiamin deficiency and possibly in scurvy. In beriberi heart disease the underlying physiologic change is peripheral arteriolar dilatation acting much like a large arteriovenous fistula associated with fatty degeneration of the myocardium. Vitamin B deficiency in infancy is not a common cause of cardiac hypertrophy in the U.S.A. although its exact frequency is difficult to ascertain. The possibility of a nutritional and vitamin deficiency should be considered in allergic infants receiving elimination diets. A fatal instance of an infant on such a diet developing beriberi associated with encephalopathy was observed by Davis and Wolf. A therapeutic trial of thiamin should result in a decrease in the size of the heart and reversion of flat or inverted T waves although this reversibility has not been noted in some advanced cases.

III. Inadequate Coronary Blood Flow

a. Anomalous Origin of the Left Coronary Artery: Several types of variation in anomalous origin of the coronary arteries have been described. A single coronary artery may arise from the aorta to supply the entire myocardium. This usually does not cause any ill effects. Anomalous origin of both coronary arteries from the pulmonary trunk is an exceedingly rare anomaly resulting in early death. Origin of the right coronary artery from the pulmonary artery with normal origin of the left coronary artery has been described in asymptomatic patients. Origin of the left coronary artery from the pulmonary artery is a rare anomaly that has been frequently described.

The clinical picture of anomalous origin of the left coronary artery resembles very closely that seen in other examples of the Left Ventricular Hypertrophy Syndrome. Feeding difficulties commence in early infancy and are followed by the development of cardiac failure. The literature contains frequent reference to angina-like attacks which are said to be peculiar to, and typical of, an anomalous left coronary artery. These attacks are precipitated by feedings or by other exertional efforts. We believe these attacks are noted in infants with myocardial ischemia from a variety of causes and are not diagnostic of this anomaly. They are not present in all patients with this anomaly and they have been observed in patients with other lesions causing the left ventricular hypertrophy syndrome. We have observed these symptoms in an infant with endocardial fibroelastosis. House reported a one-year-old child with idiopathic myo-
carditis who had anginal attacks with pain, sweating and pallor. Similar attacks have been reported by Engel in an infant with rhabdomyosarcoma. It is apparent that a history of attacks simulating angina have no specific diagnostic importance. They are of importance in suggesting the possibility of the left ventricular hypertrophy syndrome.

Upon examination, the heart is found to be markedly enlarged with primary involvement of the left ventricle. Significant murmurs are not heard. Radiologic examination reveals the presence of a large globular shaped heart. Occasionally left ventricular enlargement or aneurysmal dilatation is noted. Electrocardiographic studies of some of the first reported cases revealed inverted T waves in standard leads I and II with low voltage and ST segment deviations. These abnormalities were believed to be pathognomonic of the presence of an anomalous left coronary artery. It is now recognized that these changes are the result of myocardial anoxia and may be produced by a variety of lesions. Precordial leads often reveal deep Q waves and marked ST segment deviation suggestive of myocardial necrosis. Such changes are seen most often in lesions producing inadequate coronary blood flow and may be helpful in diagnosis. However if damage to the myocardium is sufficiently extensive from other causes, one can expect such electrocardiographic aberrations. Furthermore, abnormal electrocardiograms are not found in all cases of anomalous origin of the left coronary artery. A similar occurrence was reported by Tedeschi and Helpern. The time when the electrocardiogram is taken in relationship to the pathogenesis of this anomaly is important. In a recent experience, the first electrocardiogram demonstrated minor T waves changes. Subsequently a myocardial infarction occurred resulting in diagnostic electrocardiographic changes indicative of severe myocardial necrosis. This was confirmed by post-mortem examination. Aortography may be a useful adjunct in establishing the presence of an anomalous left coronary artery. Injection of dye into the aorta may permit radiographic visualization of the coronary arteries and their origin.

The clinical course is characteristically that of rapid progression following the onset of symptoms. In most cases death occurs in the first year of life. There are reports, however, of persons with this condition who reached adulthood and then succumbed suddenly or after progressive angina and heart failure.

At post-mortem examination the anomalous origin of the left coronary artery is noted. The more common form is the origin of the anterior descending branch from the pulmonary artery or conus while the circumflex arises from the aorta. However the entire left coronary artery may have an anomalous origin. Bland, White and Garland demonstrated that scarring of the myocardium occurs, being concentrated usually toward the endocardial side of the anterior wall of the left ventricle and septum following the distribution of the left coronary artery. There may be severe myocardial necrosis, aneurysmal dilatation of the apex of the left ventricle and, on rare occasions, rupture of the myocardium with resultant hemopericardium. Endocardial fibroelastosis involving the dilated left ventricle is a very common secondary manifestation of this anomaly.

It is of practical importance to establish, ante mortem, the existence of the abnormal origin of the left coronary artery. Surgical correction,
aimed at increasing the oxygen content of blood entering the anomalous coronary artery, has been attempted. Edwards suggests that the relative oxygen unsaturation of blood in the coronary artery is not the cause of myocardial necrosis. Low pulmonary artery blood pressure may be inadequate for maintenance of adequate coronary circulation to the left ventricle. One can anticipate continuing efforts directed towards surgical correction.

b. Coronary Occlusive Disease: The adult form of atherosclerosis or arteriosclerosis has not to our knowledge been described as a cause of heart disease in infancy but other forms of coronary artery disease do occur although they are very rare. Stryker has classified and discussed this group of lesions. Medical sclerosis of the coronary arteries is the most common form of occlusive disease in infancy. Sclerosis and calcification are found in the media and fibrous proliferation of the intima occludes the lumina of the vessels. Myocardial infarcts have resulted. No lipid deposits occur. Thrombi may be present within the lumina. Thomas reports the association of endocardial fibroelastosis with arterial calcification and myocardial infarction. In almost all cases there is generalized arterial involvement, many organ systems being affected. Coronary occlusive disease in infancy may be "idiopathic" and result in death in very early infancy. A genetic defect in the arterial elastic tissue has been suggested. Some cases are secondary to renal hyperparathyroidism due to severe anomalies of the kidneys.

The clinical picture is that noted in other lesions in this syndrome. Lipman has suggested that the ocular fundi may show evidence of arteriolar damage. Andersen and Schlesinger reported calcification of arteries in the upper extremities seen on x-ray films obtained for possible changes in the radius and ulna, while Cochrane suggests the possibility of demonstrating calcified vessels in the neck in this manner. The electrocardiogram may reveal evidence of nonspecific myocardial anoxia, left ventricular hypertrophy or of myocardial infarction.

DISCUSSION

In order to establish the presence of the left ventricular hypertrophy syndrome, it is important to demonstrate anatomic left ventricular hypertrophy in an acyanotic infant with absent or insignificant cardiac murmurs. This is not an easy task in early infancy. Displacement of the apex impulse downwards and to the left and the presence of a heaving apical thrust usually are present if significant left ventricular hypertrophy exists. A parasternal lift is usually associated with right ventricular hypertrophy. However, this is not always obvious. The generally accepted radiographic criteria of chamber enlargement are not necessarily valid in infancy. Often left ventricular hypertrophy produces a heart of rather globular shape with deviation of the esophagus posteriorly and to the right. The angiogram offers the most objective evidence of cardiac enlargement and is of diagnostic value, but we do not suggest that this procedure is necessary or advisable in all circumstances. The electrocardiogram may demonstrate evidence of myocardial anoxia or damage. Abnormalities include ST segment depressions or elevations, and T wave depressions or inversions in one or more standard and unipolar leads. Low voltage may be present. These changes are not specific for any particular anomaly but are characteristic of any disease or malformation which results in myocardial anoxia or damage. Similar electrocardiographic changes are associated with digitalis effect upon the myocardium. The myocardium may be affected by a variety of noxious agents and yet will react electrocardiographically in a uniform fashion. We do not believe the electrocardiogram has pathognomonic features. The demonstration of electrocardiographic findings in infancy simulating those of an acute myocardial infarction suggests severe disturbance of coronary circulation. These have been observed in idiopathic myocarditis,
tumors of the heart, coronary occlusive disease as well as in anomalies of the coronary arteries.

The electrocardiogram is often of value in the identification of specific chamber enlargement. Precordial tracings may reveal the presence of unequivocal left ventricular hypertrophy. Electrocardiographic criteria are available for the diagnosis of left ventricular hypertrophy. These criteria are based on the normal limits of various deflections in each age group. The finding in an infant of the typical adult R/S ratio progression from right to left-sided chest leads is strongly suggestive of the presence of left ventricular hypertrophy. The absence of the criteria necessary to establish the presence of left ventricular hypertrophy in an infant's electrocardiogram, however, does not eliminate the existence of such hypertrophy. The electrocardiogram represents the balance of forces between the two ventricles. Beyond early infancy, the normal balance of electrical forces during ventricular depolarization depends upon a left ventricle which is anatomically dominant over the right; therefore an electrocardiogram which indicates a normal balance by adult standards actually reflects some left ventricular preponderance. In order for the classical pattern of left ventricular hypertrophy to appear in the electrocardiogram, considerable hypertrophy must exist. Consequently a "normal" electrocardiogram in an infant does not exclude the presence of lesser degrees of left ventricular hypertrophy.

We must next consider the electrocardiographic diagnosis of right ventricular hypertrophy for, if one could demonstrate unequivocally the presence of anatomic right ventricular hypertrophy in a given infant, this would exclude the existence of the left ventricular hypertrophy syndrome, except in neonates. In young infants, normal physiological right ventricular preponderance exists; pathological left ventricular hypertrophy could thus be present and yet be masked for a time. As normal maturation proceeds, there is less and less anatomic right ventricular preponderance until, finally, the left ventricle predominates.

This normal development from right to left ventricular preponderance in early infancy is reflected in both the electrocardiogram and the vectorcardiogram. We have found the latter particularly helpful in this regard. In the normal neonatal state of anatomic right ventricular predominance, the vectorcardiographic pattern is that associated, in older children and adults, with pathological right ventricular hypertrophy. (In the transverse plane, there is clockwise rotation of the QRS loop and displacement of the loop to the right and anteriorly). The electrocardiogram can be predicted from the vectorcardiographic pattern, reveals tall R and small s waves in right-sided chest leads. After a period of several weeks or months, the vectorcardiogram displays a normal counterclockwise rotation of the QRS loop in the transverse plane. However, the loop is still displaced anteriorly and to the right. This displacement causes, in the electrocardiogram of infants of this age, persistence of the neonatal tall R and small s waves. At this time it may be difficult to differentiate the electrocardiogram of this "normal infant pattern" from that produced by persistent and pathological right ventricular hypertrophy. However, we believe that the vectorcardiographic configurations are characteristic and dissimilar after the neonatal period that one can easily distinguish the "normal infant pattern" from that of pathological right ventricular hypertrophy.

Electrocardiographic criteria of left ventricular hypertrophy are difficult to establish in infancy. The absence of the criteria for right ventricular hypertrophy in an infant with an enlarged heart is therefore of considerable importance. Under these circumstances we believe the vectorcardiogram is more accurate than the electrocardiogram in the first year of life.

In our experience and that of others, the axis deviation as determined from the standard leads of the electrocardiogram is of less diagnostic value. Left axis deviation occurs not infrequently in patients with right ventricular hypertrophy and vice versa.

SUMMARY

In infants a group of cardiac lesions exist which produce cardiac enlargement, predominantly of the left ventricle, without the presence of significant murmurs or central cyanosis, and in which there is a tendency to the development of heart failure. Under these circumstances the electrocardiogram may be normal, reveal evidence of myocardial anoxia or of left ventricular hypertrophy. We have called this complex the Left Ventricular Hypertrophy Syndrome. This syndrome does not include more commonly occurring cardiac anomalies which produce left ventricular hypertrophy associated with significant murmurs. It does include (1) primary endocardial fibroelastosis, (2) predominantly myocardial disorders such as (a) idiopathic or aseptic myocarditis, (b) glycogen storage disease of the heart, (c) tumors of the heart and (d) nutritional deficiency states causing myocardial dysfunction; and (3) disorders resulting predominantly in abnormalities of coronary blood flow as (a) anomalous origin of the left coronary artery and (b) coronary occlusive disease.

The demonstration of left ventricular hypertrophy in infancy by physical examination, x-ray film and electrocardiography is discussed and the usefulness of vectorcardiography in delineating pathological right ventricular hypertrophy and thereby ex-
including a diagnosis of left ventricular hypertrophy syndrome is emphasized.

The prognosis for survival beyond infancy is grave in these patients and the differentiation from congenital cardiac malformations is important.

RESUMEN

Existe un grupo de lesiones cardíacas en los infantes que producen crecimiento del corazón con predominio en el ventrículo izquierdo sin que haya murmullos de significación ni clausura central y en los que hay tendencia al desarrollo de insuficiencia cardíaca.

En esas circunstancias el electrocardiograma puede ser normal o bien revelar anoxia cardíaca o hipertrofia del ventrículo izquierdo. Hemos llamado a este complejo el Síndrome de hipertrofia Ventricular Izquierda.

Este síndrome no incluye las anomalías más corrientes del corazón que producen hipertrofia ventricular acompañada de murmullos evidentes. Incluye los siguientes: 1) Fibroelastosis endocárdica primaria, 2) trastornos predominantes del miocardio tales como a) miocarditis aséptica o idiopática, enfermedad por acumulación de glucogeno en el corazón, c) tumores del corazón, d) estados de deficiencia nutricional causantes de disfunción miocárdica; 3) trastornos resultantes de predominio de las alteraciones del flujo coronario tales como: a) origen anómalo de la arteria coronaria, y b) enfermedad coronaria oclusiva. La demostración de hipertrofia ventricular izquierda en la infancia por el examen físico, los rayos X y la electrocardiografía se discuten y se recalca la utilidad de la vectocardiografía la delinear la hipertrofia ventricular derecha y de ese modo a excluir el diagnóstico del síndrome de hipertrofia ventricular izquierda.

El pronóstico para la sobrevida más allá de la primera infancia es grave en estos enfermos y es importante la diferenciación de las malformaciones congénitas.

RESUME

Chez les petits enfants, il existe une catégorie de lésions cardiaques qui provoquent une hypertrophie cardiaque, atteignant principalement le ventricle gauche, sans qu’il y ait de souffles notables ni de cyanose. Cet état a tendance à évoluer vers l’insuffisance cardiaque. Dans ces conditions l’electrocardiogramme peut être normal, ou apporter la preuve d’une anoxie myocardique ou d’une hypertrophie du ventricle gauche. Nous avons appelé ce complexe le “syndrome d’hypertrophie ventriculaire gauche”.

Ce syndrome ne comprend pas les anomalies cardiaques beaucoup plus banales qui entrainent une hypertrophie ventriculaire gauche, mais comportent des souffles importants. Il comprend: 1°) la fibro-élastose endocardique primaire; 2°) les troubles myocardiques prédominants, tels que: a) la myocardite aseptique ou idiopathique, b) l’affection cardiaque à surcharge glycolégique, c) les tumeurs cardiaques, d) les états de déficience nutritionnelle provoquant une dysfunction myocardique; 3°) les troubles résultant de façon prédominante des anomalies du débit coronarien tels que: a) origine anormale de l’artère coronaire gauche, et b) affection occlusive coronarienne.

La mise en évidence de l’hypertrophie ventriculaire gauche dans l’enfance par l’examen physique, la radiologie et l’électrocardiographie est discutée, et l’auteur insiste sur l’utilité de la vectocardiographie pour préciser l’existence d’une hypertrophie ventriculaire droite et exclure ainsi un diagnostic de syndrome d’hypertrophie ventriculaire gauche.

ZUSAMMENFASSUNG

Bei Kleinkindern findet sich eine Gruppe mit Herzveränderungen, gekennzeichnet durch Herzvergrößerung, vorwiegend am linken Ventrikel, ohne Vorliegen nennenswerter Geräusche oder zentraler Zyanose, und bei denen eine Tendenz zum Zustandekommen eines cardialen Versagens besteht. Unter Umständen kann das Elektrokardiogramm normal sein, das Bestehen myocardialer Anoxiezeichen oder eine Hypertrophie des linken Ventrikels. Wir haben diesen Krankheitskomplex bezeichnet als "Linkseitiges Kammerhypertrophie-Syndrom."

Dieses Syndrom enthält keine der häufig vorkommenden cardialen Anomalien, die mit einer Hypertrophie verknüpft sind mit de Anomalien des Geräusches. Eingeschlossen sind (1) primäre endocardiale Fibroelastose (2) überwiegend myocardiale Störungen wie (a) idiopathische oder aseptische Myocarditis, (b) Glykogen-Speicherkrankheit des Herzens, (c) Tumoren des Herzens und (d) aginntäre Mangelszustände mit nachfolgender myocardialer Dysfunktion; (3) Störungen als Folgen besonders bei Veränderungen der koronaren Zirkulation wie (a) Anomalien im Ursprung der linken Coronararterie und (b) zu Coronarverschluss führende Krankheit.

Der Nachweis der linkssseitigen Kammerhypertrophie während der frühsten Kindheit durch physikalische Untersuchung, Röntgenaufnahme und Elektrokardiographie wird besprochen und der Wert der Vektorkardiographie zur Abgrenzung pathologischer rechtzeitiger Kammerhypertrophie mit daraus sich ergebendem Ausschluss eines linksseitigen Kammerhypertrophie Syndromes hervorgehoben.

Die Prognose hinsichtlich des Überlebens der ersten Kindheit ist ernst bei solchen Kranken und die Abgrenzung gegenüber kongenitalen Missbildungen ist bedeutsam.

References will appear in reprints.