

## THE PATHOLOGY OF CONGENITAL HEART DISEASE.\*

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Under this term may be discussed all those conditions of the heart which deviate from the normal and which date their origin between the appearance of the earliest rudiments of the heart in the embryo and the expulsion of the fetus from the uterus.

Many classifications have been suggested, but none are thoroughly satisfactory, some being too broadly comprehensive, some only a detailed enumeration of the lesions found. Thus, one classification may be based on etiology, the anomalies being due to: 1, faults of development; 2, fetal endocarditis; 3, a combination of both, either one preceding the other and predisposing to it. Other classifications may be based on the morbid anatomy, on the symptomatology or on the prognosis. For the purposes and limits of this paper, the one already given will perhaps answer best, with a brief working-out along certain lines.

*Faults of Development.*—A combined anatomic and chronologic division of these may be made into three classes: 1, those occurring early, from the fourth to the sixth week, showing a heart with two or three cavities, *cor biloculare* or *triloculare*, with a single or imperfectly divided arterial trunk; 2, anomalies arising between the sixth and twelfth weeks, with imperfect auricular or ventricular septa, imperfect or misplaced vessels; 3, defects occurring after the twelfth week, resulting in anomalies of the valves, persistence of fetal opening, etc.

*Fetal Endocarditis.*—Other changes than those already mentioned are usually the result of fetal endocarditis. This arises often as the result of some infectious process in the mother, especially rheumatism, but any infectious fever may cause it; syphilis in the mother is said to play an important part. It may also occur without evidence of illness on the part of the mother. Whether it is necessary, in order to produce the endocarditis, for the germs of disease themselves to pass through the placenta from the maternal to the fetal circulation, an event which certainly happens in some processes, or whether the products of bacterial activity, the toxins circulating in the maternal organism, may by a process akin to dialysis enter the fetal blood, can not be stated in every case. Probability in favor of the latter view is strengthened by the form of endocarditis which is invariably the sclerotic or chronic and never the warty or verrucose. The right heart is far more frequently the seat of the inflammation than the left. The usual reason adduced to explain this is the greater amount of work done, as, in extrauterine life, the left side is more frequently affected. It has been suggested, however, by Rosenbach, that the richness of the blood in oxygen is the determining factor. Another reason given is that the right heart is more often the seat of congenital anomalies of the valves, and these are predisposed to inflammation, just as in extrauterine life diseased valves are a weak spot and prone to recurrent inflammation. Heredity is of great importance according to some authorities, in favoring both anomalies of development and fetal endocarditis, as shown by Mousous' classic cases.

With reference to the anatomic lesions, the following table from Holt's work is interesting, the order given being that of the frequency with which the lesions were present in 242 cases: defect in the ventricular septum; defect in the auricular septum or patent foramen ovale; pulmonary stenosis or atresia; patent ductus arteriosus; abnormalities in the origin of the great vessels; pulmonary insufficiency. The most frequently associated lesions were: pulmonary stenosis with defect of the ventricular septum; pulmonary stenosis with defect of the auricular septum; defects in both septa; pulmonary stenosis with defects in both septa.

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## THE DIAGNOSIS OF HEART DISEASE IN CHILDREN.\*

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This paper has been prepared on such short notice as to give me no opportunity to consult medical journals and text-book literature. What I have to say is, therefore, purely the result of my own experience with affections of the heart in children.

The first question which naturally arises in diagnosis is, is heart disease present, and if so, is it congenital or postnatal?

Let us take up first the study of congenital heart disease. The principal diagnostic symptoms of this condition are: cyanosis, clubbing of the fingers, thrill, characteristic murmurs, the absence of any great enlargement of the heart.

Cyanosis is peculiarly marked in congenital heart disease; in fact, I do not know of any other condition in which it is so intense. Even in severe forms of postnatal heart disease, with entire lack of compensation and decided blueness of the lips, I never have seen the blue-ered tongue, the purplish cheeks, and the general blue suffusion of the body present in the congenital cases. This form of cyanosis, then, is alone an important diagnostic symptom.

Clubbing of the fingers, when present, is a very characteristic symptom. It occurs, it is true, in chronic disease of the lungs, but even the worst cases in this condition show clubbing no greater than, if as great as, that seen in congenital heart disease. Combined, therefore, with cyanosis, clubbing of the fingers is of great diagnostic importance.

The thrill of congenital heart disease is very characteristic. It is very intense, rough, and widely diffused. One may strongly suspect the existence of a congenital affection of the heart from the character of the thrill alone. Yet it is by no means always present.

The typical murmurs of congenital heart disease are loud, rough, and of great intensity. The intensity, in fact, is out of all proportion to the other physical signs connected with the heart. The situation of the murmurs, too, is peculiar. They are not commonly heard with greatest loudness at the apex, but rather over the base of the heart, the sternum and the aortic and pulmonary cartilages. It must be remembered, however, that even in young children there may be loud basic murmurs dependent solely on the existence of great anemia. We must be careful, therefore, not to make the diagnosis from the character of the murmurs alone. I

\*Read in a Symposium on Heart Disease in Children, before the Philadelphia Pediatric Society, April 10, 1900.

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shall not go into any extended discussion of the diagnosis of the nature of the individual valvular affections, from the character and position of the murmurs, further than to say that those present in pulmonary stenosis are heard loudest at the pulmonary cartilage, while those due to a perforate septum ventriculorum have their greatest intensity over the middle of the sternum. We must remember, too, that the presence of cyanosis in conjunction with a murmur points strongly toward the existence of stenosis at the pulmonary orifice, while the existence of cyanosis without any murmurs awakens a suspicion that there is complete closure of the pulmonary orifice or an anomalous origin of the great vessels.

The fact that there is commonly no great enlargement of the heart in congenital heart disease is peculiar. At most there occurs some hypertrophy of the right side. The condition is in striking contrast to the very great enlargement which is so commonly seen in cardiac affections developing after birth.

The second question is, is there a postnatal heart disease? We must consider *a*, the value of the symptoms apart from the physical signs, and then *b*, the diagnostic value of the physical signs themselves.

*a*. It must be stated at the very outset that the symptoms of heart disease in childhood are quite commonly insignificant or absent altogether. Children have a peculiar power to acquire perfect compensation of a valvular defect. Consequently, heart disease in children is frequently discovered purely by accident, there often being no subjective symptoms. When such symptoms do exist, chief among them is to be mentioned shortness of breath. Very often the presence of this can be elicited only by careful questioning of the child or of the parents. Sometimes we learn that the little boy can not keep up with his companions in active games, or we find him listless and disposed to sit around and play at quieter games with his girl companions. In my experience, the existence of slight shortness of breath is quite commonly the only symptom. In more advanced cases, we may have the ordinary intense dyspnea characteristic of heart disease at any age. In estimating the value of shortness of breath as a diagnostic symptom, we must remember that anemic children may readily exhibit this symptom to a decided degree.

Edema is a suspicious symptom. It quite commonly begins in the feet and it develops also in the abdomen. It is very important, however, when making a diagnosis, to remember that children become edematous quite easily under certain conditions. We have, for instance, the edema of post-scarlatinal nephritis, which, not at all infrequently, appears first in the feet and not first in the face, as we usually expect it to do. Then there is the edema of the feet which so often develops in marantic infants. Next, the edema dependent on grave anemia is not uncommon. Again, we must remember that ascites in children is not uncommonly due to a tubercular peritonitis, and not to a heart affection. I have at the present time under my observation one or two cases in which the diagnosis between a heart disease and a tubercular peritonitis is by no means certain. Finally, I have repeatedly seen in children a curious tendency to the development of marked and widespread edema without the existence of any discoverable cause whatever.

Faintness is a symptom occasionally seen in children with postnatal heart disease, but in my experience, it is not at all common. I recollect very well one child whose first symptom was a tendency to faint after he had

climbed the stairs. Only careful study showed that the disease in this little boy was in reality a cardiac affection.

Precordial pain is in my experience not common in children. I have seen it marked only in advanced cases with extreme lack of compensation. In these I have sometimes found the pain very severe.

Palpitation is a symptom which is of very little diagnostic value in childhood, not so much that it is not present, as because children do not recognize the palpitation or do not know how to describe it. I have seldom found this symptom present.

Cough is in childhood a symptom of no great diagnostic value in cardiac affections. I have found it only where other much more prominent symptoms of the disease were present. In these cases it was an evidence of intense passive congestion of the lungs.

The presence of a decided degree of anemia is sometimes a diagnostic symptom of considerable importance. Many children who have heart disease for some time have a distinctly delicate and anemic appearance which seems to be the direct result of the cardiac affection.

I have found hemoptysis uncommon except in bad cases. The tendency to it, which is not at all uncommon in mitral stenosis in adult life, has seemed to me to be decidedly rare in childhood. I have come across it but very seldom.

*b*. Next, we have the consideration of the diagnostic value of the physical signs connected with the examination of the heart itself. The first question which arises naturally is, does the existence of murmurs prove the presence of heart disease? This is an old question to which the well-known answer is "no." The fact that anemia may be attended by very well marked murmurs in childhood as well as in adult life is well understood and has been already referred to. We have also the existence of what may be termed accidental murmurs, the origin of which we do not understand, but which can not be associated with any certain disease of the valves of the heart. Again there is the murmur which not infrequently develops during and disappears after fever. There are also certain organic murmurs produced outside of the heart, which simulate very closely the endocardial murmurs which are the subject of the present discussion. I refer to the murmur of pericarditis and the cardiopulmonary murmur which has often been described.

On the other hand, heart disease may exist, as we very well know, without the presence of any murmurs whatever, or the murmurs may perhaps come and go for reasons which we do not well understand.

In connection with the study of the heart murmurs, we have two other matters to be carefully studied also: the pulmonary second sound and the size of the heart.

With regard to the pulmonary second sound, we know that its accentuation is a common sign of disease of the mitral valve. In childhood, however, we must carefully remember that the second sound is physiologically accentuated; that is to say, in early and later childhood we expect to find the pulmonary second sound normally as loud as, or even louder than, the aortic second sound. No one can with certainty make a diagnosis of heart disease in childhood without bearing this fact in mind.

Then, with regard to the size of the heart, hypertrophy and dilatation as results of valvular affections become extremely marked in childhood, more so than in adult life. There are certain factors, however, which render the diagnostic value of this sign sometimes uncertain. For

instance, the presence of a nephritis will naturally produce enlargement of the left side of the heart. On the other hand, an asthma with the consequent emphysema will cause the lungs to overlie the heart and make its size appear less than it really is. Then, too, we must remember that in childhood, before the age of puberty, the right side of the heart reaches somewhat farther toward the right than it does in adult life, and that the apex-beat is quite commonly in the fourth interspace instead of the fifth. If these facts be not borne in mind, one might readily make the diagnosis of hypertrophy of the right ventricle when no such hypertrophy was really present.

If, while bearing in mind the various diagnostic symptoms mentioned and the questions which arise in connection with them as already pointed out, we have reason to believe that postnatal heart disease exists, the next matter to be decided is, what variety is it? I can not here enter on this subject further than to say that in mitral insufficiency we expect to have enlargement of both ventricles, but particularly of the right, with very great accentuation of the pulmonary second sound. If one fails to find the accentuation of this second sound, in accordance with the age of the child, the diagnosis often remains somewhat uncertain.

In the case of mitral stenosis, the right side of the heart becomes enlarged, but usually not the left, and the pulmonary second sound is very greatly accentuated. Quite commonly in this affection we find a very marked thrill, which is diastolic in time. The murmur, too, is diastolic, thus distinguishing mitral stenosis from mitral insufficiency, which is systolic in time. With both these lesions, when compensation fails, one naturally gets the symptoms of pulmonary congestion. Other forms of heart disease are less common in childhood, and comments on their symptoms and physical signs must be omitted.

In general, it may be said that in diagnosis of postnatal cardiac affections in childhood we should observe the following points: avoid making a diagnosis from the presence or nature of murmurs alone; remember the altered position of the right side of the heart and of the apex-beat in childhood; remember that the presence of an accentuated pulmonary second sound is normal, not pathologic; remember that compensation is acquired very easily in childhood, and that the absence of symptoms does not prove the absence of cardiac disease; remember that the most suggestive symptom is dyspnea, and that edema must be studied most carefully before it becomes of value as a diagnostic symptom.

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## REPORT OF A CASE OF PRIMARY CARCINOMA OF LIVER.

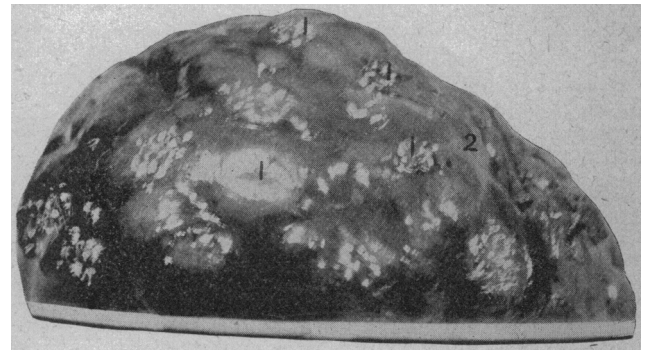
BY R. C. HARRIS, M.D.

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Robert B., aged 57, a farmer, and native of Switzerland, entered the St. Louis City Hospital March 3, 1899, giving the following history: He had always enjoyed good health, suffering only with the minor ailments of childhood, and with an attack of double pleurisy at the age of 20. His family history was negative. He was accustomed to drinking on an average, five glasses of beer daily, and a small one of whisky early in the morning. He had smoked a pipe incessantly from 12 years of age. He denied having had syphilis, and has never been injured. The malady that ultimately caused his death began about two years ago, at which time, while laboring in the field he was suddenly seized with an exer-

ciating pain in the pit of the stomach, the right side of the chest and the right shoulder, which compelled him to stop his work and rest. After a few moments the pain lessened, but a dull, dragging sensation came on. A physician who was summoned pronounced it a case of gall-stone colic, blistered over the site of pain, also gave cathartics and hypnotics. The patient made a slow recovery, the pain entirely leaving him, but the dull dragging sensation, "the feeling of weight," as he expressed it, mentioned above persisted in the region of the gall-bladder. Soon he noticed that if he attempted to walk fast, or exerted himself to the slightest degree, he would grow tired, dizzy and dyspneic. His ankles began to swell, the swelling extending gradually up the limbs, finally invading the abdominal cavity. His physician withdrew, by paracentesis, about a gallon of bloody ascitic fluid, giving the patient considerable relief.

From this time on he enjoyed fair health, suffering only with the dragging sensation mentioned above, until Feb. 1, 1899, when he began to suffer with a reiteration of the first attack. On entering the hospital, the following were the conditions: The patient was exceedingly well nourished, weighing about 225 pounds, not the slightest trace of jaundice or emaciation being present. The pupils were extremely dilated, but reacted to light and distance normally. The temporal arteries were tortuous and sclerotic, the tongue coated and flabby, and there were visible systolic pulsations in the jugulars. Examination of the chest revealed harshened respiratory murmur of both sides, also a few piping râles over the base of both lungs posteriorly. The apex-beat was about an inch to the left of the nipple and in the fourth interspace. A



1. Cancer Nodules. 2. Thickened Peritoneum.

soft systolic murmur was heard over the mitral area, not transmitted in any direction; a similar one over the tricuspid area, this being plainly audible all over the sternum and right upper chest.

Further examination showed the abdomen to be large, rounded, and capped by a prominent caput Madusa. Several small petechial spots were seen about the neighborhood of the umbilicus. The lower portion of the abdomen was occupied by fluid, determined by succussion, percussion and aspiration. The upper part was occupied by a hard mass, which did not move with respiration. This "mass" extended down to about 1½ inches below the umbilicus; its lower border was rounded and smooth, except in one spot directly over the gall-bladder, where a single hard nodule was found, and which was exceedingly painful to the patient when pressed upon. This was about the size of a small walnut. A peculiar fremitus was elicited by palpation over this area, not unlike the peculiar fremitus of hydatids, but thought in this case to be a localized perihepatitis. Percussion showed the growth to extend upward as far as the fifth rib on the right side; its lower border became imperceptibly lost on the left side under the free margins of the ribs. I succeeded, by the use of the trocar, in withdrawing about a gallon of a sero-sanguinous fluid, which microscopically showed the presence of leucocytes, pigment and blood, the latter being demonstrable by the Teichman hemin crystals.

Uranalysis showed: specific gravity, 1010; straw-colored; acid reaction; slight trace of albumin by heat, nitric acid