SPECIAL ARTICLE

Approach to Diagnosis of Congenital Heart Disease without Recourse to Special Tests

By Alexander S. Nadas, M.D.

The title of my presentation of course, raises the question: What is a special test? I would think that I am expected to discuss procedures customarily performed in the office today. One can take a good history and do a good physical examination. One can take x-rays, do fluoroscopies, and electrocardiograms.

The next point I would like to raise concerns the use of some of these office tools such as the stethoscope. I would like to put in a word that you should use a stethoscope of the proper length (10 inches), with proper tubing (1/8-inch bore), equipped with 2 end-pieces, a diaphragm, and an open bell. I also must tell you that I have found surprising improvement in the accuracy of my auscultation by doing the very complicated maneuver of cleaning out the ear pieces.

Next I would like to make a few remarks about a second tool, the fluoroscope. Fluoroscopy can teach us a lot, but it involves a great deal of radiation, relatively speaking. My radiologist friends tell me that a minute of fluoroscopy under ideal circumstances, generates about 6 r's, whereas only .25 r is involved in a posteroanterior film of the chest. So I would like to caution you to use fluoroscopy sparingly. In our hospital we fluoroscope all patients initially, and then no more often than once every 2 to 3 years, and in the majority, even less frequently, when specific reasons arise. We rather prefer to gage the progress of the patient with 7-foot posteroanterior films, and possibly oblique views. Our radiologists think, and I know that not everybody agrees, that the pulmonary vasculature may actually be judged better by films than by fluoroscopy. This, then, is an added reason for the use of films.

As far as electrocardiography is concerned, I don't think I have to spend time on general advice. I would like to say only that if you are dealing with children, please don't be satisfied with the usual V1 to V6 leads, but take V1*, maybe V2*, and leads above or below V4u, so that you get a complete R/S progression from a dominant complex on one side of the chest to a dominant complex on the other. We have been using vectorcardiography to a certain extent within the past year, and it is proving to be a fairly useful addition, giving information not obtainable by the conventional scalar leads in certain specific instances.

Now once we are equipped with all these tools, I would like to ask, "What is really the role of the clinician in evaluating congenital heart disease?" The first question I have to answer when I am confronted with a child—this being 90 per cent of my experience—is, does he have heart disease, or not? This is very important, since we are all confronted in our daily practice by a number of children referred with a tentative diagnosis of heart disease, who, on the basis of a thorough office examination may be pronounced healthy. I would like to propose certain criteria for the diagnosis of heart disease in children.

I would like to use the type of classification that Dr. T. Duckett Jones used for rheumatic
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fever, and say that there are certain major and minor criteria. By major criteria I mean, for the purposes of this discussion, that the presence of any one of them should enable the clinician to make the diagnosis of heart disease (table 1). The first thing, of course, is a systolic murmur more than grade III in intensity. If this is accompanied by a thrill, then the diagnosis of heart disease can be even more certain; but even without a thrill, a grade III (on a scale of I to VI) systolic murmur is usually adequate evidence on which to base the diagnosis of heart disease.

The second major criterion is a diastolic murmur of any intensity. This alone is proof positive, in the vast majority of instances, to establish the diagnosis of heart disease.

The third point is congestive heart failure. This, even in the absence of a murmur, proves the presence of heart disease. Of course, one has to look out for pitfalls, and I hasten to assure this group that we have fallen into these ourselves, just as much as anybody else in this audience. We have had a patient with "nephrosis" sitting on our ward for a month before right-sided congestive failure was recognized. We also have had small babies with "asthma," who, on investigation, turned out to have left-sided failure. On the whole, however, signs or symptoms of left or right-sided failure, per se, are enough to establish the diagnosis of heart disease.

The final major criterion is cyanosis. Again, you have to watch out for pitfalls. Abnormal hemoglobins, pulmonary disease, well-water methemoglobinemia and all the other things one tortures medical students with at examination time ought to be thought of, and usually discarded. If you can be sure that the cyanosis is indeed cardiac in origin, you may make a diagnosis of heart disease with assurance.

The second group of criteria, the minor ones, are those of which 2 are needed for a certain diagnosis of heart disease. Among them is a systolic murmur less than grade III in intensity. These are the faint murmurs, and many may be classified as innocent. Some-

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<thead>
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<th>Major 1</th>
<th>Minor 2</th>
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<tr>
<td>Systolic murmur &gt; III</td>
<td>Systolic murmur &lt; III</td>
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<tr>
<td>Diastolic murmur</td>
<td>Abnormal x-ray</td>
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<td>Congestive heart failure</td>
<td>Abnormal electrocardiogram</td>
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<td>Cyanosis</td>
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thing else such as an abnormal electrocardiogram or x-ray must go with it before the diagnosis of heart disease can be established.

But an abnormal electrocardiogram alone is again probably insufficient evidence to make the diagnosis of heart disease. Within the framework of coronary disease, this may not be true; but in children, I think the electrocardiogram, although we have good standards for normal, is by itself probably not enough to establish the diagnosis of heart disease. On the other hand, if you couple the electrocardiogram with another criterion, it becomes decisive.

Abnormal x-rays may give us a great deal of trouble, particularly in small babies with a large thymus. They may have large mid-thoracic shadows. The parents live in fear and trepidation for months, saying, "This child has heart disease." In fact, the heart may be normal, but the thymus is large. Another pitfall in this regard is the difference in heart size between inspiration and expiration films; I am sure this is known to everybody. Still it may be well worthwhile to stress this point in order to reinforce the principle not to accept an abnormal x-ray without other criteria as convincing evidence of heart disease.

I would also like to say a few words about the abnormality of the second sound at the second left interspace. Sometimes the degree of the splitting, or the intensity of the second sound, may be a very important clue to lead you to recognize or discard heart disease. A widely split second sound, particularly if it does not change with respiration, suggests an atrial septal defect. A markedly accentuated pulmonary closure indicates pulmonary hypertension. A diminished second sound means,
usually, low mean pulmonary arterial pressure.

The final criterion is the abnormality of blood pressure, particularly the relative hypertension of the upper extremities characteristic of coarctation of the aorta. This may be used almost as a major criterion, but I doubt that you will find it without the appearance of murmur or abnormality of the electrocardiogram or x-ray.

Once we have established the diagnosis of heart disease on any basis, the next point is, "Is this congenital or is it acquired?" In children I think this is fairly easy. Obviously there is the time factor. Although people talk about rheumatic heart disease appearing congenitally, or within the first year of life, I believe that this is quite rare in the experience of those who see many patients with rheumatic heart disease. If a patient has a murmur or other evidences of heart disease under the age of 1, or even under the age of 2, you may assume that it is congenital in nature.

It should be emphasized, however, that the late discovery of a murmur is not necessarily evidence against congenital heart disease. There are certain types of congenital cardiac defects, particularly ostium secundum atrial septal defect, in which murmurs are particularly prone to appear or to be discovered late. The murmurs of an atrial septal defect are commonly discovered for the first time during an examination for entering school. Coarctation of the aorta, with the relative hypertension of the upper extremities and the murmur over the back, is just as commonly discovered when the child is 10 or 12 years old. Finally, although a patent ductus arteriosus is discovered quite often in infancy, we see a surprising number of youngsters who have had good pediatric care in whom the characteristic Gibson murmur does not appear, or is not discovered, until 3 or 4 years of age.

It may be worthwhile to contrast the conditions in which murmurs usually appear early in life. Foremost among these are aortic stenosis, pulmonic stenosis, and ventricular defect.

In addition to time factors, the presence of cyanosis means congenital heart disease for all practical purposes. Then there is the location of the murmur. Parasternal systolic murmurs suggest congenital disease strongly. Conversely, apical murmurs do not mean rheumatic heart disease, necessarily; many congenital conditions manifest themselves by a loud murmur at the apex.

If on the basis of the foregoing principles one has established the diagnosis of congenital heart disease, we may turn our thoughts toward one or another specific entity with the aid of certain group characteristics.

The first large group to think of is the group of so-called left-to-right shunts. In these patients there is a communication between the systemic and pulmonary circuit anywhere from the patent ductus down to the atrial level, including pulmonary venous anomalies. The group characteristics of the left-to-right shunts include (1) pulmonary vascular engorgement by x-ray; (2) hyperkinetic cardiac impulse; (3) left-sided chest prominence; (4) lower left sternal border, or apical proto- or mid-diastolic, so-called "shunt," rumble; and (5) absence of cyanosis. If the diastolic rumble is loud, if the pulmonary vasculature is not only engorged, but also shows expansile pulsations, and if the cardiac silhouette is appreciably enlarged, one may assume that the shunt is of considerable magnitude.

The next big group to be considered is the groups of right-to-left shunt. These are the patients with cyanosis and clubbing. Occasionally, these 2 features may not be present in equal degree. The cyanosis may be minimal, or may only be represented by an intense redness of the toes and fingers, but the clubbing may be marked. This large group of cyanotic patients may be subdivided into those (a) associated with pulmonary stenosis (stenotic, systolic murmur at the second left interspace, diminished pulmonary closure, and ischemic lung fields); (b) those with pulmonary vascular obstruction (pulmonary ple-
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thor, loud pulmonary closure, and slight systolic murmur); and finally (c) the trans-
positions (underdevelopment, pulmonary ple-
thora, and absent main pulmonary artery at the usual place).

Finally, the third large group of congenital heart diseases are the obstructive lesions with-
out shunts, such as coarctation of the aorta, aortic stenosis, and pulmonic stenosis. These
patients, as a group, are characterized by a diamond-shaped stenotic, systolic murmur of-
ten accompanied by a thrill. The cardiac impulse, in contrast to the left-to-right shunt
process, is heaving. The presence of chest de-
formity is relatively rare, and diastolic rum-
bles are usually absent or faint. Cyanosis is
of course not present.

Once one has classified the patient into one
of these large groups, then on the basis of individual features, an accurate diagnosis may
be rather easily made. For instance, in the obstrusive group, if the murmur is loudest
over the back, one probably is dealing with a
coarctation of the aorta. If it is best heard
at the second left interspace, it is probably
pulmonic stenosis, whereas with aortic stenosis
the same type of murmur is heard to the right
of the sternum. Within the left-to-right shunt
group, in a similar fashion, a secundum-type
atrial defect may be recognized by the wide
and fixed splitting of the second sound, and
a soft, ejection-type systolic murmur at the
second left interspace. By contrast, an endo-
cardial cushion defect is identified by a mean
electrical axis of between $-20$ to $-180^\circ$, and
a ventricular defect is recognizable by a harsh,
systolic murmur at the lower left sternal
border. Of course, the ductus can easily be
identified by a continuous machinery murmur.

All of this, of course, is not simple, and
particularly the cyanotic group may be ex-
tremely complicated. I still believe, however,
that with the intelligent use of the eyes, ears,
and hands, the electrocardiogram, and the
x-rays, the diagnosis of congenital heart dis-
ease may be made with relative ease in about
75 per cent of the cases. I also think that
probably about one third of the remaining
25 per cent cannot be accurately diagnosed
by any means short of an autopsy.

All of this does not lead me to say not to
use all the tools available for accurate ana-
tomic and physiologic diagnosis necessary to
give a maximal amount of information for the
surgeon; but I do mean that with an intelli-
gent use of the clinical tools, one can direct
the physiologic studies into the more fruitful
avenues, and may gradually arrive at the
point where a good number of these lesions
can actually be accurately analyzed without
the use of more complicated, cumbersome, and
expensive methods.

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