SYMPOSIUM ON CONGESTIVE HEART FAILURE

Pediatric Aspects of Congestive Heart Failure

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In most respects congestive failure is similar in all age groups, but some of the special features pertaining to infants and children are important. Detailed reviews on the subject have been published within recent years.1-3

One of the most significant differences between the congestive failure of adults and children is an etiologic one. Adults develop heart failure usually on a rheumatic, arteriosclerotic, or hypertensive basis. By contrast, congenital and rheumatic heart diseases are the principal etiologic groups in pediatrics; it is our impression that congenital heart disease leads more commonly to congestive failure than rheumatic heart disease, but no good statistical support of this view is available. Other less frequent etiologic entities in childhood are primary myocardial disease, paroxysmal supraventricular tachycardia, acute glomerulonephritis, anemia, and pericarditis.

We shall not discuss all these entities in detail but rather point briefly to some of their unusual features. We shall also discuss the clinical picture of congestive failure in children in general and make a few brief remarks about therapy and prognosis.

Congenital Heart Disease

It is important to realize that the myocardium of children in congestive failure due to congenital heart disease is usually relatively healthy. Consequently, children represent a therapeutically more hopeful group in which successful surgical repair of the lesion may improve the circulation sufficiently to allow normal life expectancy.

The congenital cardiovascular abnormalities most commonly leading to congestive failure are in order of frequency, transposition of the great arteries, coarctation of the aorta, ventricular septal defect, aortic atresia, common atrioventricular canal, transposition of pulmonary veins, single ventricle, and patent ductus arteriosus.1

The age at onset of failure is noteworthy. Keith1 stated that 90 per cent of the children who develop congestive failure do so within the first year of life. This may serve as a basis for optimism to parents of children who had no failure within this period of time.

Children with the syndrome of the tetralogy of Fallot seldom, if ever, develop congestive failure. The reasons are probably the relatively small pulmonary blood flow, the normal systemic flow, and the right ventricular pressure in the systemic range. We have so far encountered only 2 circumstances that led to the development of congestive failure in Fallot's tetralogy; namely, bacterial endocarditis and severe anemia. The latter may be a so-called "relative anemia" with hemoglobin values within the low normal range for children without arterial unsaturation.

Critical pulmonic stenosis with intact ventricular septum resulting in high right ventricular pressures sometimes twice the systemic level may, by contrast, lead to severe congestive failure early. The auscultatory findings of pulmonic stenosis without severe cyanosis, marked cardiac enlargement, and severe right ventricular hypertrophy by electrocardiogram may suggest this diagnosis. Emergency surgical intervention may be lifesaving.

Severe coarctation of the aorta may lead to congestive failure in early infancy. If failure with this malformation does not occur within the first 6 months of life, it rarely occurs before the second decade. Surgical treatment for
alleviation of congestive failure should be employed only if vigorous medical management fails. Successful anticongestive measures may foster adequate growth and development and a return of the heart size toward normal. Relief of the aortic block then may be postponed to a later optimal time.

Among the groups of left-to-right shunts the ostium secundum atrial defects almost never lead to congestive failure in infancy and very rarely in childhood. By contrast some of the children with the severest congestive failure are those with an ostium primum septal defect. The addition of mitral and tricuspid regurgitation to the hemodynamic load of the atrial defect is probably the reason for this difference between the 2 types of atrial defects. Large ventricular defect with pulmonary arterial hypertension and a large pulmonary flow is also likely to lead to congestive failure early in infancy. Medical management usually, though not invariably, tides these infants over to an age when surgery can be performed with relatively low risk.

Patent ductus arteriosus is about the only left-to-right shunt leading to congestive failure that at the present time should be corrected surgically as soon as the diagnosis is made. Medical management should be utilized only as a preparation to operative intervention.

Rheumatic Heart Disease

The appearance of congestive failure on a rheumatic basis is very rare under 2 years of age and probably assumes numerical significance only beyond 5 years. It has repeatedly been stated that rheumatic fever in the young is usually accompanied by maximal evidences of carditis. An additional axiom, the origin of which is hard to trace, is that a child with congestive failure on a rheumatic basis always has active carditis. One further point to be stressed is that children with congestive failure due to rheumatic heart disease always have a murmur; it may be stated with assurance that if no significant apical murmur is heard, the etiology of congestion must be other than rheumatic.

Other Etiologic Entities

Arrhythmia

An analysis was made of the incidence of failure in children with arrhythmia. Cardiac decompensation was present in 22 of a group of 41 children, 29 of whom had supraventricular tachycardia,4 9 had atrial flutter, and 3 had ventricular tachycardia. Of those with failure, 75 per cent were 4 months old or less. Failure was not seen if the arrhythmia lasted less than 24 hours but was present in 19 per cent of those in whom it lasted 36 hours and in 50 per cent of those in whom it persisted for 48 hours. The heart rate, once it was above 180 per minute, seemed to have little influence on the development of failure; as many children with failure had heart rates from 180 to 240 as from 250 to 330.

Paroxysmal supraventricular tachycardia was almost always terminated successfully with digitalis. In the rare refractory case propranolol was used with success.

Myocarditis

One of the most significant recent contributions in the field of heart disease of the young has been the isolation of Coxsackie virus group B from the hearts of infants succumbing from cardiac failure early in life.5-8 The disease has appeared as early as 13 hours after birth and as late as 7 weeks of age. The illness has been characterized9 by the onset of difficulty in feeding, fever, lethargy, and progressive cardiac failure. Less than half of the reported infants had symptoms of the central nervous system or hepatosplenomegaly. Twenty-one of the 27 patients have died.9-11 The remaining 6 seem to have recovered completely.

Postmortem examinations revealed patchy necrosis and inflammation of the myocardium; the left ventricle was affected more than the right. Lesions have also been described in the brain, meninges, liver, spleen, adrenal and pituitary glands, pancreas, bone marrow, and striated muscle.11

In many cases a history of mild febrile illness, accompanied by headache or pleural pain has been elicited from the mother or
The virus has been isolated on many occasions from contacts of the infant. Intrauterine\textsuperscript{9} as well as postnatal infection with this agent has been postulated and nursery epidemics have been reported.\textsuperscript{5-7} The difference in susceptibility of the host to the virus is striking; the disease is mild in adults, but usually fatal in the newborn.

Two cases have also been reported of children with myocarditis and associated pericarditis. The virus was isolated from both patients who recovered.\textsuperscript{10, 12}

Some of these babies have been treated with corticosteroids in addition to the usual anti-inflammatory measures. The authors are not convinced of the superiority of this additional therapeutic effort.

In the majority of the myocarditides in childhood no etiologic agent can be identified, and they are termed idiopathic or Fiedler’s myocarditis. There is little doubt that with the improvement of diagnostic techniques specific etiologic agents will be identified. In the meantime all should be treated with vigorous, though cautious, anti-inflammatory regimens.

Chronic extensive myocardial fibrosis, sometimes familial in occurrence, in other instances following an acute infection, often simulates the picture of constrictive pericarditis in children.

**Acute Glomerulonephritis**

In acute glomerulonephritis acute pulmonary edema is a rare complication but the onset may be sudden with rapid progression to a catastrophic climax. Quick and decisive action by the physician in face of such an emergency may prove to be lifesaving. Treatment should include oxygen, possibly under positive pressure, intravenous rapidly acting digitalis compound, aminophylline, morphine, and possibly tourniquets. Pulmonary edema may be the presenting symptom of acute glomerulonephritis with or without systemic arterial hypertension.

**Anemia**

Severe anemia may lead to congestive failure. Hemoglobin levels below 5 Gm. per 100 ml. of blood caused by leukemia, Mediterranean anemia, sickle-cell anemia, hypoplastic anemia, or even simple iron-deficiency state may cause severe cardiac failure even if the heart is structurally normal. Slightly higher levels (less than 7 Gm. per 100 ml.) may contribute to the development of congestive failure if the heart muscle is damaged by rheumatic carditis or nephritis or if structural deformities are present.

It is a good rule not to transfuse these children with whole blood but rather with increments of packed cells, slowly (5 to 10 ml. per pound every 8 to 12 hours). We have also tried in some infants with severe anemia to remove small aliquots of blood, replacing it with equal amounts of packed cells, thus avoiding an increase in circulating blood volume. The use of digitalis has not been particularly effective in the treatment of congestive failure secondary to anemia.

**Pericarditis**

Pericarditis as a cause of congestive failure is rare. Rheumatic pericardial involvement usually denotes severe pancarditis, and the accompanying failure is due to severe myocardial involvement. Constrictive pericarditis is very rare in the pediatric age group. Despite clinical and physiologic indications of this diagnosis, surgical exploration almost invariably reveals a normal pericardial sac. At autopsy, extensive myocardial fibrosis is usually found.

Acute purulent pericarditis, secondary to pneumonia or bacteremia, may cause pericardial tamponade and congestive failure. Rare instances of so-called acute "benign" pericarditis or severe rheumatoid arthritis also may lead to cardiac tamponade. Finally, in the postoperative period after cardiotomy, hemopericardium may represent a serious surgical emergency.

Pericardial tamponade causing severe rightsided failure is particularly difficult to identify in infants whose short necks do not allow adequate evaluation of the jugular venous pulse. The discovery of marked cardiac enlargement and muffled heart sounds without murmurs, coupled with a high index of suspicion in the conditions mentioned, may lead
to the consideration of pericardial effusion in the differential diagnosis. Careful search for the presence of pulsus paradoxus of over 10 mm. of mercury, low systolic and narrow pulse pressure, clear lung fields, and electrocardiographic changes of pericarditis may lead to the correct diagnosis. Pericardial tap or pericardial exploration under local anesthesia, if necessary, is the procedure of choice if the diagnosis is seriously suspected. We wish to stress the inadequacy of radiologic and even fluoroscopic attempts to distinguish between a maximally dilated heart and accumulation of fluid in the pericardial sac.

Once the diagnosis of cardiac tamponade is made, surgical evacuation of the compressing fluid is the treatment of choice. If the fluid is purulent, vigorous treatment with antibiotics is indicated. Digitalis usually is not helpful, although we cannot attest to its being harmful. Diuretics may be indicated.

Clinical Manifestations of Congestive Failure

The clinical picture of congestive failure differs to a certain extent in children from that commonly observed in adults. Purely left-sided failure with paroxysmal nocturnal dyspnea, rales at the bases, frothy blood-tinged sputum, and the "butterfly" distribution of pulmonary edema on the radiogram is practically unknown in children. Contrariwise, the clinical profile of right-sided failure with facial edema, hepatomegaly, and jugular venous distention is the common picture. Periorbital edema was so marked in a 20-month-old girl with atrial tachycardia of 1 month's duration that she was sent to the hospital with the diagnosis of nephrosis. Other manifestations of right-sided failure, such as dependent edema and ascites, are found less often in children than in adults. Anorexia, irritability, excessive perspiration, and restless sleep are common to both left- and right-sided failure.

Although the dominant feature of heart failure in the pediatric age group is systemic congestion, most of the patients indeed represent combined right- and left-sided failure, with the former dominating the picture. Although the evidences of systemic congestion are obvious, left-sided failure may be suspected only by observing tachypnea in a patient without cyanosis. Heart failure in even purely left-sided lesions (patent ductus arteriosus, coarctation of the aorta) is often recognized first on the basis of peripheral edema and hepatomegaly; the primary left ventricular failure is manifested only by the more subtle sign of tachypnea. Sometimes mothers note rapid respiration before the appearance of systemic congestion. This is often not recognized by physicians as an indication of left-sided failure.

Purely right-sided failure is seen in relatively few instances: in pure pulmonic stenosis, in pulmonary vascular obstruction, in complete transposition of the pulmonary veins, and occasionally in atrial defect. Purely left-sided failure is seen rarely in some infants with a large patent ductus, severe coarctation of the aorta, endocardial fibroelastosis, or mitral valve obstruction.

### Table 1

<table>
<thead>
<tr>
<th>Dosage of Oral Digitalis</th>
<th>0 to 2 years</th>
<th>2 to 13 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total digitalizing dose given in 3 divided doses</td>
<td>0.03 to 0.04 mg./lb.</td>
<td>0.02 to 0.03 mg./lb.</td>
</tr>
<tr>
<td>Daily maintenance dose given in 2 divided doses</td>
<td>1/4 to 1/3 digitalizing dose</td>
<td>1/4 to 1/3 digitalizing dose</td>
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The therapy of congestive failure in a child differs little from that in an adult. The general medical principles used in treating adults also apply to infants and children.

Any one of several preparations of digitalis can be used in pediatrics with benefit. A suggested dosage schedule for Digoxin is given in Table 1. The principle of individualizing digitalis dosage to achieve maximal therapeutic or minimal toxic effect cannot be overemphasized. Unusual sensitivity to digitalis may be present in some patients with myocarditis, so that in them digitalization should be accomplished cautiously under close observation.

Diuretic agents are safe as well as effective.
in the young. Tables 2 and 3 list approximate dosages according to age. The intravenous administration of aminophylline one half hour after a mercurial injection also is useful in children who do not respond adequately to mercurials and digitalis alone. Considerations regarding electrolytes and diuretics in the pediatric age group and in adults are similar.

A low-sodium diet should be given only if it can be made palatable enough for the child. Children need an adequate protein intake for growth. Consequently, if the only way a qualitatively and quantitatively adequate diet will be consumed is by offering foods with normal sodium content, we would choose to do so if more diuretics have to be given in order to maintain an edema-free state. If, on the other hand, a low-sodium diet is tolerated and the need for diuretics can be reduced or abolished, this course would seem preferable.

The physical comfort of the child and relief of restlessness and anxiety are extremely important. Morphine has proved to be an invaluable drug in this regard. Doses of 0.05 to 0.075 mg. per pound are usually satisfactory. Similar doses are also sufficient for the treatment of acute pulmonary edema. A device that we have found useful for propping young babies in an upright position is illustrated in figure 1. If desired, it can be converted easily from a seat to a flat surface so that the infant may be examined in the supine position. An oxygen tent may be used if it does not frighten the child and is kept cool, but it should quickly be abandoned if the baby is afraid of it.

Since many infants with rales have pneumonia as well as congestion of the lungs, antibiotics are probably indicated if there is any sign of infection or if the child is debilitated. In the latter case fever and leukocytosis may be absent in the presence of pneumonia.

**Summary**

Congestive failure in infancy and childhood may occur in a number of diseases. Congenital heart disease, however, is probably the most common cause of failure, with rheumatic fever second. Although the basic physiology of cardiac decompensation is similar in adults and children, the presenting clinical picture may be somewhat different in the 2 groups.

We believe that infants and children with congestive failure should be treated vigorously, for we have the distinct impression that the mortality rate of these youngsters with severe heart disease has decreased appreciably since a more optimistic attitude prevails among parents and doctors about these problems. Vigorous use of anticongestive measures and antibiotics is worthwhile and results in many instances in the survival of an infant.
to an age when successful surgical repair is feasible. Certainly not all babies with congestive failure due to congenital, rheumatic, or other kind of heart disease can be salvaged. But surely the experience of the last few years has given us a much more optimistic outlook on this entire field. There are enough children attending school and enjoying life today who seemed at “death’s door” in infancy to make us hesitate to give an unqualifiedly poor prognosis to almost any child in congestive failure. Similarly, we are more and more reluctant to suggest cardiac surgery as a dramatic emergency measure except in a few well-chosen cases. Intelligent medical management can salvage a large number of these children and allow them to survive to an age when relatively low surgical mortality may be offered.

**Summario in Interlingua**

Congestive disfallimento cardiae potest occurrere durante le infantes et pueritia in association cum un numero de morbos. Tamen, congenite morbo cardiae es probablemente le causa le plus commun de disfallimento, con febre rheumatic occupante le secunde rango. Ben que le physiologia fundamental de discompensation cardiae es simile in adultos et juveniles, le aspectos clinic de presentation pote differer in un certe maniera in le 2 gruppos.

Nos opinque que infantes et juveniles con disfallimento congestive debe esser tractate vigorosamente, proque nos ha distinctemente le impression que le mortalitate in iste gruppo de patientes pediatric ha decresee appreciablemente deposit que un attitude plus optimista predominant le partis de parentes et medicos con respecto a iste problemes. Le uso vigorose de mesuras anti-congestive et de antibiotics vale le pena et resulta in multe casos in le superviventia de un infante usque al etate ubi le reparo chirurgic pote esser effectuare in un successo. Certo, il non es possibile salvar omne le infantes con disfallimento congestive resultante de morbos cardiae congenite, rheumatic, o altare. Sed nonobstante il es un facto que le experientias del passate annos justifica un prognose multo plus optimista in iste campo integre. A iste tempore un sufficentemente grande numero de juveniles visita le scholas e gaudi del vita ben que illes se trovava “in mano de morte” in lor infantea pro non permitter a nos le formulation de un uniformemente sombre

**References**
