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Heart Failure in Infancy and Childhood *

by

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Abstract.

Recognition of heart failure, especially left heart failure is often more difficult in the infant than the medical management. Treatment of failure is aimed at;

1. *Rapid relief of congestion in vital organs such as the lung and*
2. *Improving the oxygen supply to deprived organs.*

Basically the plan should be aimed at increasing cardiac output, decreasing circulating blood volume and preventing conditions which call for increased oxygen utilization.

For patients with pulmonary edema attention is directed at improving ventilation as much as in improving cardiac output.

Heart failure in the infant may now be treated with greater optimism and hope than was justified ten years ago. Some infants recover entirely with no further treatment needed while others recover sufficiently to reach the cardiovascular surgeon in a reasonable state for definitive surgical repair.

* Invited paper at the Third National Indonesian Paediatric Congress, Surabaya, July 1 - 6, 1974.

Received 30th. Nov. 1974.

Three — fourth's of patients with cardiac disabilitity in infancy have some form of cardiac failure. Recognizing this it is surprising to consider that over 50% of the babies referred to our hospital with cardiac failure were first treated for some non-cardiac condition assumed to be the cause of the complaints for days, weeks or months before the basic problem was recognized. As obvious as heart failure, sometimes it is difficult to understand why there is this problem in diagnosis.

Part of the difficulty stems from the terminology, "congestive heart failure". If there is no congestion to be found on examination, that is, no liver enlargement nor audible rales in the lung, many physicians are unwilling to make the diagnosis of heart failure. This is a technicality which is dangerous to the health of the patient.

Traditionally the heart is failing when either it is unable to provide adequate flow to the organs and tissues of the body or when extraordinary means of compensation are required to provide adequate oxygen delivery. The pure physiologists would have us to understand that heart failure means a measurable reduction in cardiac output. While in heart failure secondary to myocardial disease the cardiac output is indeed low, in many of the congenital malformations of the heart the output measures higher than normal simply to compensate for the extra load of the defect.

If we are to expand the concept of heart failure to include the congenital cardiac defects in an effort to improve early diagnosis, we must think of heart failure in another way. The heart may fail in the task which the defect is imposing no matter how unreasonable these demands may be. For example, the heart may fail to accept all of the increased filling arriving at a specific chamber resulting from a large left — to — right shunt. In other anomalies one or more of the chambers may be required to build up an unreasonably high pressure to overcome obstruction to the outlet of the chamber.

Under these circumstances the heart, by the mechanism of hypertrophy and increased contractility may manage to pay its oxygen debt to the body tissues. This compensation might prove effective for a considerable but unpredictable time before the circulatory system must resort to some of the extraordinary means of achieving oxygen delivery such as tachycardia, a widened arterial venous oxygen difference or an increased ventricular end-diastolic pressure. Under these conditions while the oxygen delivery is accomplished the adaptations required to do so are not without detriment to the well — being of the patient.

The purpose of identifying heart failure in a patient is to commence

promptly with appropriate management and to alert the clinician to the ominous nature of the disease. The designation of "failure" usually, but not always, calls for digitalis and diuretic treatment. Furthermore, it suggests the need for definitive anatomic diagnosis at a selected time and place. The diagnosis of congestive heart failure in a patient with a surgically treatable anomaly should bring up at least a consideration of the need for surgical treatment.

If the purists wish to reserve the term "heart failure" for those circumstances in which there is inadequate cardiac output to meet oxy gen requirements, we might compromise by identifying what the immediate precursors are to this state of unequivocal heart failure. Then we could strive to treat the patient who manifests these warning signs, with all of the care and concern sometimes reserved for the individual with each of the traditional and for advanced indices of cardiac decompensation.

In the congenital malformation of the heart what are these precursors and what are their clinical equivalents?

1. Left atrial overload.

Anomalies such as large ductus arteriosus or ventricular septal defect cause an increase in return of pulmonary venous blood to the left atrium. If the atrial septum is intact this volume of blood must be contained within the left atrium during ventricular systole while the mitral valve is closed contained within the left atrium or else refluxed back into pulmonary veins. One of the experimentally confirmed dynamic characteristics of the left atrium is that excessive filling volume within this chamber causes a sharp rise in pressure. In this respect the left atrium differs from the right atrium which has its own built - in escape routes for volume overload both by way of the vena cava and also the more easily dilatable walls of the right atrium. The pulmonary veins have muscular walls near their entry into the left atrium and it would appear that this initially protects the vulnerable pulmonary veins from a mild elevation of left atrial pressure rise, but only to a limited extent. Very soon the full thrust of left atrial hypertension is reflected back into the pulmonary veins. This leads then to ultimate exudation of fluid at the pulmonary capillary level with all of the consequences of pulmonary edema, ventilatory insufficiency and pulmonary arterial hypertension.

The patient first breathes faster in order to improve gas exchange. When fast breathing becomes inadequate he begins to breathe with extra respiratory muscles and must build up a high intrathoracic pressure to move enough air for vital ventilation. When this increased intrathoracic pressure become quite

high the pulmonary capillaries may close. Insufficient blood passes through to the left heart and a weak pulse results following every exaggerated breath — pulsus alternans may appear to be present, actually a strong respiratory variation. This so-called paradoxical pulse may simulate that seen in pericardial tamponade.

Infants who must work so hard to breathe use extra calories and fail to gain weight properly; of course, poor weight gain is to some extent the result of the inability of the infant to take feedings well. Furthermore, this extra work of breathing causes the infant to sweat excessively. He becomes very fatigued and this undoubtedly interferes with his appetite and makes him often a very irritable baby. The excessive sweating sometimes produces a sudaminous rash which is itchy and certainly does not improve the infant's disposition.

Pulmonary edema causes a cough which is often presumed to reflect a respiratory tract infection especially in the infant who has none of the obvious manifestations of heart disease. But if rales in the lung are not heard pulmonary edema may not be suspected even in the infant who has a known heart anomaly. In the United States treatment for a "cold" often includes antihistamines. This medication however helpful for relieving symptoms of a "cold" has the adverse effect of drying secretions which may obstruct terminal bronchioles and lead to segmental atelectasis and pneumonia.

Why is it that infants with pulmonary edema seldom have audible rales on clinical auscultation? A certain velocity of sufficient air flow is necessary for rales to be heard. When we examine the adult we ask him to breathe deeply to bring out the rales. In the infant with an inherently low tidal volume to begin with, reduced further by pulmonary edema and by inadequate lung compliance and fatigued respiratory muscles, a reduced volume of air moving at a slow rate will not produce the sound we recognize as rales despite the presence of interstitial or intra-alveolar edema. A simple corollary is the powerful blast of breath necessary to produce a sound thru a trumpet.

The radiographic appearance of pulmonary edema is a subtle finding and may be missed, especially if the film is overexposed. The radiographic features are also often misinterpreted as pneumonia especially if the radiologist is provided with that clinical impression. When the heart is dilated on the chest roentgenogram the correct diagnosis of pulmonary edema is supported by that finding but in lesions which obstruct the return of pulmonary venous blood to the left atrium the heart size may remain normal and this

makes recognition of pulmonary edema very difficult.

Left atrial and pulmonary venous hypertension is exaggerated by a number of events which are likely in the life of the infant. These include exercise such as feeding or crying, being placed in the supine or prone position and stress situations causing an increased flow through the heart such as fever, infection or anemia. Pulmonary venous hypertension may be aggravated by an intravenous fluid infusion or worse yet an ordinary blood transfusion. From the physiologic standpoint growth alone brings on an increase in circulating blood volume which exaggerates the problem. Once reduced output develops and fluid retention increases the circulating blood volume the problem becomes further amplified.

Conditions which interfere with emptying of the left atrium likewise lead to elevation of pressure with all of the resulting symptoms and signs just discussed. There are numerous situations in which this might occur. One is simply an elevation of the left ventricular end — diastolic pressure resulting from inadequate left ventricular contractility or in situations in which the healthy left ventricular myocardium has been excessively overloaded by a congenital defect. The site of obstruction to left atrial emptying may be at or near the mitral valve and there are numerous anomalies which can produce this. Left ventricular overload whether from increased filling volume or severe increase in pressure leads to the same type of symptoms and signs in the infant as mentioned for left atrial overload.

If the clinician identifies left atrial or left ventricular dilatation by whatever means — physical examination, chest roentgenogram or electrocardiogram, this finding could be taken as an adequate warning for the potential development of pulmonary edema whether all of the other manifestations mentioned are present or not.

Overload of the right heart.

Overload of the right heart is a different matter. As mentioned earlier, increased volume returning to the right atrium raises right atrial pressure very little and less abruptly than is characteristic of left atrium. The pressure is dissipated by right atrial dilatation and there are no symptoms from this event per se. Great dilatation of the right atrium especially in older children and adults predisposes atrial tachyarrhytmias but except for this complication, so long as output of the right heart is adequate very little symptomatic discomfort is felt by the patient or noted by the parent simply from right atrial dilatation. The same is true when right atrial pressure rises in some of the severe obstructive lesions causing interference

with right atrial emptying. Likewise, symptoms are few and late in anomalies causing increased volume of blood filling the right ventricle or in those leading to increase in pressure. When right heart output is interfered with, either by decrease in heart rate or decrease in right ventricular contractility or progressive increase in degree of mechanical obstruction, then symptoms may appear and these have to do with an abrupt decrease in filling volume of the left side of the heart and consequently to the systemic circulation. Then symptoms of fatigue, lightheadedness and even syncope with effort may occur. But these patients are remarkably comfortable at rest, maintain a good appetite and continue to gain weight. They do not sweat excessively, are not orthopneic nor irritable until late in the course of events.

The findings of right heart overload appear earlier or perhaps are easier for the clinician to detect since the liver enlargement is somehow a simpler thing to recognize on physical examination than is rapid breathing characteristic of left heart overload.

The consequences of precursors of heart failure.

Right heart overload is much less devastating than left heart overload as seen in the congenital cardiac defects. Left heart failure has its greatest effects on ventilation, it exhausts the patient and leads to early hypoxemia and hypercapnea. But congestion of the liver and peripheral edema is less serious to vital organ function. Long standing right heart decompensation leads to protein loss in the intestinal tract and after many years, to hemosiderosis of the liver. However, the ability of the right heart to vent itself by back pressure into the systemic venous system without direct consequences to the patient allows the right ventricle, overworked as it may be, to maintain an adequate resting output for a long time. What the right ventricle may not be able to do is to meet the sudden demand of some acute stressful situation which is why apparent "sudden death" is known to occur in this type of derangement.

Differences in pediatric and adult age in manifestations of heart failure.

Differences occur in pediatric and adult age patients in the manifestations of heart failure, some of which have already been mentioned. In addition the opportunity for congestive heart failure is greater in the newborn infant. There are several reasons for this :

1. The reactive pulmonary vascular bed of the neonate predisposes to an abrupt rise in pulmonary artery pressure in response to hypoxemia. Furthermore, this neonatal phenomenon accounts for persis-

tence of pulmonary hypertension secondary to volume overload resulting from large left to right shunts.

2. Renal immaturity adds to the reaction in renal blood flow and glomerular filtration rate.

3. Infant is likely to suffer fatigue of the respiratory muscles more easily from the work of breathing than the older child or adult.

4. Functional metabolic derangements characteristic of the newborn who is stressed from any cause include hypoglycemia, hypocalcemia and hypokalemia. These electrolyte and metabolic derangements contribute to an exaggerate heart failure.

5. Hepatic handling of lactate is relatively inadequate in the infant.

The other differences between the infant and the adult have to do with such elementary, but often overlooked, factors as :

1. The inability of the infant to express his needs in terms understood by the parent, the nurse and the physician. When the infant feels uncomfortable he cries but infants also cry for a number of other biologic needs. It requires very astute and sensitive attention to the infants needs to determine if his crying is anything more than physiological. In infants with breathing discomfort due to pulmonary edema, when the parent or nurse picks the baby up and places him in an up right position this lowers pulmonary venous pressure simply by gravity and partially relieves his discomfort. The infant thus may stop crying and the insensitive observer may conclude that the baby was crying for attention.

The consequences of inadequate cardiac output.

There are several organ and tissue responses to a reduced cardiac output.

1. Cutaneous arterial vaso — constriction.

One of the earliest compensations the body makes to a reduction in cardiac output is cutaneous ischemia. The more vital organs take priority for the blood flow available and by the phenomenon of redistribution, cardiac reserve is called upon causing the skin arteries to constrict. This produces a skin pallor and may elicit a false clinical impression of anemia. Since there is usually hemodilution from the increased circulating blood volume secondary to low output, the hemoglobin may, in fact, measure less than normal. If a false diagnosis of anemia is made and treated with oral iron, little harm is done other than delaying the diagnosis but a blood transfusion under these circumstances may prove disastrous.

2. Renal arterial vasoconstriction.

Reduced renal function occurs early with reduced cardiac output, leading to increased circulating blood volume and the loss of protein in the

urine. The appearance of red blood cells and white blood cells in the urine may further suggest primary renal disease. This false diagnosis may be further supported by the appearance of facial edema which in infants may develop earlier than edema of the extremities.

3. Gastrointestinal arterial vaso — constriction.

Redistribution of blood flow may involve the gastrointestinal tract to provide better flow to the more vital organs .As a result, varying degrees of ileus develop with subsequent intestinal and gastric distention which interferes with feeding and compromises diaphragmatic motion. Patient in whom gastrointestinal symptoms predominate and who lack the obvious signs of a heart defect may find themselves being investigated for some form of intestinal disease.

4. Cerebral ischemia.

Little deprivation of cerebral blood flow occurs by a mechanism of redistribution. The brain is the vital organ which is spared by the process of redistribution of blood flow from the skin, gastrointestinal system and kidneys. However, a systemic arterial mean pressure falls late in the course of severe left ventricular failure, then hypoxia occurs and cerebral symptoms do appear. These consists of drowsiness, stupor and ultimately coma. But many patients succumb from other sequelae of heart failure long before cerebral symptoms occur. In newborns, however, cerebral hypoxia symptoms from reduced left ventricular output may mimic brain damage from other causes and many patients with a severe congenital heart defect who lack a murmur or other obvious signs of heart disease undergo a neurological investigation.

5. Heart failure begets heart failure.

Renal ischemia leads to fluid retention and the increased circulating blood volume exaggerates the already overload heart and lungs. Low systemic flow produces tissue hypoxia leading to accumulation of lactates. These products of anoxic metabolism are not readily eliminated both because of renal insufficiency and hepatic insufficiency, a particular problem in the young infant. Lactates lead to acidemia rendering the body's catecholeamines relatively inactive. Thus, myocardial contractility is further impaired.

Management of heart failure.

Management of heart failure is usually simpler and is carried out by the clinician than is early recognition. There are, however, a few recent advances in the medical treatment of congestive heart failure:

1. Management of heart failure now can be carried out in the infant

with congenital heart disease with a great deal more optimism than in the past. For one thing, new diuretics are more effective; for another prompt medical treatment allows many infants to reach a cardiac center and thus arrive in a diagnostic unit or in the operating room in better condition. Furthermore, effective medical treatment of congestive heart failure in some infants allows them to survive long enough to benefit from the chance of spontaneous closure of the defect.

This occurs in approximately 30% of patients with ventricular septal defect and in a large number with ductus arteriosus and not infrequently in atrial septal defect.

Therapy of congestive heart failure whether right or left sided is aimed at three general areas :

1. Increase the cardiac output with the use of digitalis, avoiding excessive depression of the heart rate.

2. Decrease the circulating blood volume with the use of diuretics avoiding excessive electrolyte loss.

3. Decrease the need for a higher cardiac output by reducing fever, treating infections, giving small feedings and supplying oxygen, avoiding oxygen toxicity in the newborn.

Special problems of left heart failure.

For the patient in pulmonary edema additional measures may be needed :

1. Improve ventilation by placing the infant in the upright position.

2. Avoid depression of respiration.

3. Relieve anxiety and improve tidal volume by the administration of very small doses of morphine (for example, 0.01 mg/lb of body weight). The often quoted dosage 0.1 mg/lb is clearly excessive.

4. Rapid acting diuretics

Now that diuretics such as furosemide have proved so effective it is rarely necessary to perform phlebotomy and the efficiency of these diuretics make it less important to proceed with the risky business of giving increasing doses of digitalis to near toxic levels.

5. Ventilatory support.

For extreme pulmonary edema unresponsive to these measures, endotracheal intubation and positive pressure oxygen may prove life saving.

6. Peritoneal dialysis.

In the newborn with generalized edema in whom diuresis cannot be achieved because of renal immaturity the use of peritoneal dialysis for one or two days may be very effective until renal function is established.

The management of acidemia associated with heart failure.

It is now popular to administer sodium bicarbonate to raise the blood pH in infants with severe aci-

demia. Indeed this treatment may be necessary as an emergency measure to resuscitate the patient with bradycardia which is unresponsive to atropine administration or as an urgent means to treat the patient with severe hypotension which is unresponsive to alpha stimulation with phenylephrine. But bicarbonate therapy is usually unnecessary if the acidemia has not rendered catecholeamines ineffective. Bicarbonate therapy has undesirable side effects:

1. It is only temporarily effective perhaps for five to ten minutes depending upon the severity of the cause.

2. Bicarbonate therapy leads to hyperosmolality resulting in the intravascular space attracting a intravascular space attracting fluid. This, of course, only further overloads the already overloaded circulation.

3. Recently some studies have indicated that the excessive sodium produced from bicarbonate therapy may lead to intracranial hemorrhage in the newborn infant.