Heart Disease in Infancy

Uptodate

With Dr Jane Somerville, Institute of Cardiology, University of London.

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<**Opening titles**>

< **Somerville to camera**>

During the last 10 years there has been increasing interest in the problem of heart disease in the infant.

<**Somerville narrates over statistical chart, uses indication stick**>

Now, in this country and in most countries in the world, heart disease in children is caused mainly by congenital heart disease. The remaining 10% is rheumatic, primary myocardial or due to a dysrhythmia. So if we are to concentrate on heart disease in childhood and particularly in infancy, it is the congenital heart disease that we must look at.

Now, a long time ago, in 1953, a survey by the Birmingham group, which was mainly prospective, recounted all the infants that they recognised who had had congenital heart disease and to their astonishment, 34% of these were dead by 1 month and 60% were dead by 1 year.

<**Somerville to camera**>

There is no reason, even with growing knowledge, to disbelieve this statistic. Indeed it may be very much worse than that and now that more and more infants in the neo-natal period are being recognised with heart disease and serious heart disease, one knows this to be a true fact. Thus if one was to do anything about this it was in the first year of life that one had to concentrate on.

<**Somerville narrates over graph, uses indication stick**>

Now, this slide also reflects about the age group of patients who present in infancy. I have taken, in a year, patients who were admitted to Sick Children, Great Ormond Street, as acute cardiac emergencies. On the bottom line, is the age of these infants – 2 weeks, 1 month, 3 months, 6 months. And here are the numbers of patients seen. And you see that the majority of infants with heart disease are admitted before the 3 month period. This slide was made in 1965 and if one remade it now I think there would be more falling into the 1 month period because of earlier recognition in obstetric hospitals, etc. I have also put on this slide in the cross-hatching, those that had correctable lesions, that is lesions that could be totally corrected in infancy or in whom a palliative operation was possible and later on one visualised the possibility of correction.

Now, you will see that a large number of these sick infants were able to have some corrective procedure and this shows how important it was therefore to recognise the illness and diagnose and treat it.

Now I mentioned that this slide was made in 1964 or 65 because I didn't include transposition – we were doing operations on transposition but there was a relatively poor successful return. Now if you add transposition into this, you will put up each column of correctable lesions, leaving very few lesions that we can do things about.

<**Somerville to camera**>

Thus there is a return for our interest and much can be done. Now, how does cardiac disease cause illness in the infant?

<**Somerville narrates over slides, uses indication stick, camera intermittently returns to her**>

Basically, illness results from 3 main sorts of problems. The first is, there is over-filling of the lungs with blood plus there is the association of high pressure in the pulmonary artery. Next there is obstruction causing heart failure – if it's left-sided constriction the patients will get pulmonary venous congestion and present like those with over-filled lungs. If it's right-sided obstruction, the problem will be different And lastly, babies get into trouble because of hypoxic problems, because they are blue, and we will speak about that later.

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Now let us look at the presentation and the lesions and what happens in this group of cases. Now the sort of lesions that give rise to what one might call heart failure with "stiff" lungs. Firstly they may be due to high pulmonary blood flow with high pressure and these are the large septal defects. The commonest is some form of ventricular septal defect, a large ductus, a truncus, a big A.V. canal, also transposition but that has special hypoxic problems in addition. And lastly, total anomalous pulmonary venous drainage. All these patients, their problem is basically this. Now the same clinical presentation, namely with heart failure and the 'stiff' lungs may result as I've said from pulmonary venous congestion or left-sided obstruction. And this is due to coarctation, mitral value disease, cor triatriatum and if you get obstructed anomalous pulmonary veins.

Now how does this present to the doctor? I have labelled this group with heart failure in infancy as 'plethoric lungs' – the patients with V.S.D., duct or those with congested lungs. They come because they are underweight, they are breathless on feeding and they have multiple chest infections. Unfortunately there are still babies in welfare

clinics who are underweight and having their milk changed when really they have heart failure, and this was much more of a problem a number of years ago. So this is how they will present, as really thriving problems.

Now how is one to recognise that there is an important, underlying, heart disease? Because the presence of a murmur may draw attention to the fact there's heart disease but it doesn't necessarily mean it's serious. Equally, some serious heart diseases do not have murmurs. So how are we to recognise that there is heart failure or so-called 'stiff' lungs, either due to congestion or high flow in pressure. What other signs do we look for?

Well, if the baby is tachypnoea with rates of 50 or 60 perhaps when they're asleep or supposedly at rest, or if there's a persistent tachycardia; if the chest is blown out, if there is cardiomegaly and if there is hepatomegaly.

<**Somerville reads from notes and narrates to camera**>

These five signs may lead you to suppose that perhaps there is heart disease of importance. Of course if there's a murmur it instantly draws attention to the possibility. So if you have an underweight baby with these signs, one should recognise the possibility of serious heart disease. Now you cannot recognise cardiomegaly clinically in an infant and for this reason, if there is any suspicion, such babies should be referred instantly for a chest x-ray and an electro-cardiogram.

Now this bulging of the chest is an interesting sign.

<**Somerville narrates over slide of boy's chest in profile**>

And here is a little boy and you see here is the smooth anterior bulge of the front of his chest where it has been blown out by the 'stiff' underlying lungs and there is this intercostal recession which is usually symmetrical and this occurs because the major muscle of respiration is the diaphragm, attached in this area, and it pulls hard and with the lungs being stiff there's bulging out here and pulling in here. This is a very

good sign that something important is wrong with the lungs; it also occurs in lung disease but it's a very important sign if associated with heart disease.

<**Somerville narrates over slide of boy's chest from front**>

Here you see, looked at from the front, this bilateral sulcus. This is not due to primary sternal abnormality, this is simply due to underlying 'stiff' lungs. And if the lesion is not corrected in infancy, this chest shape will be fixed as ossification occurs. If some correction is done in infancy then this chest may not develop as a permanent deformity.

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<**Somerville to camera**>

We have already mentioned that a chest x-ray is vital in these infants. And in these patients who present with what one might call a so-called 'stiff' lung / heart failure syndrome […]

<**Somerville narrates over chest x-ray, uses indication stick**>

[…] here is a chest x-ray which may confirm the cardiomegaly, seen here. You may see pulmonary plethora or increased pulmonary blood flow. This is difficult to see in the neonate but after the first 2 weeks it should be obvious. We also, as a routine, take a lateral chest x-ray because this gives important information, and you see here the bulge that we have noted clinically – here is the diaphragm pulling on the lower part of the chest and it also confirms the large heart. But another reason for taking the chest x-ray is that sometimes these patients have pulmonary oedema. And pulmonary oedema is best seen in the lateral view of the chest x-ray of an infant. If we could concentrate on this x-ray for a moment, you see the signs of pulmonary oedema well shown in this x-ray and you notice that the lines, the so-called septal lines that we normally see in the costophrenic angle, in the infant are seen anteriorly.

Now, the presence of pulmonary oedema is really an emergency and in infancy, something must be done. There are other signs of pulmonary oedema also shown and as you see, fluid in the fissure. Now, in some of the conditions that cause pulmonary oedema, the heart may not be enlarged and here you see a heart of near-normal size in the AP view; but the lungs here show the oedema although they don't show the septal lines. And here is the fuzziness of the cardiac border, the air bronchogram or the air alveologram where there is air-containing lung surrounded by oedematous lung – all important signs of pulmonary oedema.

<**Somerville to camera**>

Now I'm just going to summarise the sign of pulmonary oedema on the x-ray in infants […]

<**Somerville narrates over table**>

[…] but unfortunately it often isn't recognised and they tend to be diagnosed as obscure lung disease. Fuzzy cardiac borders, the air bronchogram, fluid in the fissures, septal lines on the lateral and the heart is often, although not always, of normal size.

<**Somerville to camera**>

And just to recapitulate, the causes of pulmonary oedema in the infant, because it's relatively uncommon I've mentioned […]

<**Somerville narrates over table**>

[…] them here. Perhaps the commonest one we see is obstruction of anomalous pulmonary veins or anomalous pulmonary veins diving below the diaphragm to enter the portal or inferior caval system. Cor triatriatum, the third membrane in the left atrium, mitral stenosis, hypoplastic left heart (which is the commonest cause of

pulmonary oedema in the neonate probably), coarctation as a sole lesion may occasionally do it and sometimes aortic stenosis.

Thus, you will see that these babies present with the ‘stiff’ lung syndrome but they may initially have small hearts and when these babies get large hearts they are usually terminal and […]

<**Somerville to camera**>

[…] therefore it emphasises the importance of recognising this syndrome and not diagnosing it as lung disease.

Turning away from pulmonary oedema let me just mention […]

<**Somerville narrates over tables, uses indication stick, camera intermittently returns to her**>

[…] possible diagnostic clues at the bedside in the baby with the ‘stiff’ lung / heart failure syndrome.

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If they have bounding pulses your first thought should be the presence of a persistent ductus, rarely an A.P. window. The continuous murmur in the infant is often heard in the left sternal edge and not out in the duct area as it is in older children. If you have bounding pulses plus a questionable cyanosis, your thought should be of a truncus arteriosis. You always feel the femoral pulses on examination, if they are delayed or absent then coarctation is present. If the femoral pulses are coming and going – one minute you think they’re there and the next you don’t, and when the baby is slightly hypoxic and cross, you can feel the pulse, when it’s quiet and in oxygen you can’t , your first thought should be that you are dealing with a pre-ductal coarctation. If you’ve got the ‘stiff’ lung / blown chest syndrome plus severe cyanosis, transposition

is an obvious diagnosis. If you have the signs of atrial septal defect and you recognise a split second sound, which you can in these infants, total anomalous pulmonary venal drainage is your first diagnosis. And if you've got a baby who has no murmurs but who appears to have heart failure and a bulged chest and is sick, consider coarctation, obstructive total anomalous pulmonary venal drainage, even transposition if they're cyanosed and of course if they are terminal, the output may not be such to allow you to have a murmur. If they look rather greyish and are low output plus a continuous murmur plus the signs of pulmonary hypertension with a loud ejection sound in the pulmonary area, once again you think of this diagnosis, particularly in the neonate.

<**Somerville reads from notes and narrates to camera**>

I have not mentioned the signs of the hypoplastic left heart syndrome, but if you see a grey baby deteriorating quickly with a rapidly enlarging heart in the first 10 days of life, I think that should be your first diagnosis.

Now having said we recognise the problem, what about treatment?

<**Somerville narrates over statistical chart, uses indication stick**>

One should say at this point that as soon as the problem is recognised, it should be sent to a specialised centre basically not because the most important thing is treatment, but because the exact anatomical cause is the most important. And the diagnosis must be made by a special investigation.

The principles of treatment are enumerated here. These babies are worried babies and they need sedation. The most active thing a baby does is feeding and this is what they can't do because they're too breathless so we use tube feeding and we put down a fine tube so the nursing staff cannot feed the baby too quickly, however rushed they feel. And these fine tubes, with a little feeding and often, these babies can be fed without causing distress. Temperature control is important – they must not be over-heated, the tendency is to swaddle the baby particularly when it's transferred

from hospital to hospital, so you have a sort of thing like a boiled baby arriving; it's a disaster for the baby in heart failure. But of course temperature control in the neonate is important so it doesn't drop its temperature. Digitalis must be prescribed for heart failure – in these babies it's a very useful drug and they take it in much bigger doses than weight for dose in adults. Diuretics may be useful, we are using Flexyx or used to use Neptal but in the neonate one must be careful when administering diuretics. Treat infections, most important, they often have concordant respiratory infections, urinary infections and these must be looked for. And of course anaemia may need to be corrected by transfusion if this is a problem; one hesitates to do this but sometimes one can do it at the time of diagnostic catheterisation because it is important, particularly if they're down in the 50s. However, as I have said already, this is very important and this is not my brief today to discuss cardiac catheterisation, etc.

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Now, having shown you medical treatment, we've diagnosed the cause, what possibilities are there for surgical treatment, because this is really the thing that has really revolutionised the outlook of these rather pathetic babies.

Now, ductus's should be ligated if they're causing heart failure; coarctation resected – there is an incidence of re-coarctation particularly if it's resected before the age of 3 months, but then the baby's life has been saved and it's justifiable. These large left and right shunts in pulmonary hypertension, there is a case for restricting the pulmonary artery and creating pulmonary stenosis. Total anomalous pulmonary venous drainage may be corrected in 1 or 2 stages, and truncus I think it's difficult to know what to do but probably there's a place for banding.

<**Somerville reads from notes and narrates to camera**>

Now, I perhaps have made this field look a little simple but in fact one of the problems of this heart disease in infants is that the lesions are very often multiple. And you may only be able to treat one to get the baby out of heart failure. But this is

worthwhile. But remembering that these lesions are often multiple, we should therefore be careful in our search for these lesions.

Now, before finishing I would like to mention hypoxic heart disease in the infant. I should remind you that there are other causes of cyanosis in the baby, particularly in the neonate.

<**Somerville narrates over tables, uses indication stick, camera intermittently returns to her**>

And these non-cardiac causes of neonatal cyanosis of course cause problems in diagnosis. Atelectasis, respiratory distress, pneumonia and pneumothorax, diaphragmatic hernia; now these present as blue babies and one sees them referred in as with heart disease. It may be very difficult to distinguish but usually a quick chest x-ray tells you the answer. A clinical clue is that if cyanosis dates from the moment of birth it's a very strong possibility that it's due to a non-cardiac cause, if it is truly from the moment of birth and stays since birth.

These are the other problems that one has to look very carefully for, but these are relatively rare. Peripheral cyanosis in the neonate is common and usually one can distinguish with careful observation.

Now, looking at the cardiac causes of cyanotic disease in the infant. Cyanotic heart disease tends to be rather complex and I've tried just to pick out the ones that one will see.

In the blue baby, of course the question of blue baby, in particular the neonate is influenced by the level of the haemoglobin, and the higher the haemoglobin is the more cyanosed the baby will look, and one has to be careful of that, particularly in the first day of life. The presence of clubbing is a useful sign of confirming the presence of central cyanosis. It is first seen in the thumb and that is the place to look first. It is rarely seen before the age of 5 weeks. But it is good to look for it and it confirms that you are dealing with a cyanotic problem.

Now, here are the conditions which we do see in the infant. By far the commonest perhaps is transposition of the great arteries and Fallot. These are relatively rare. Let us look at the problem of transposition of the great arteries.

<**Somerville narrates over statistical graph uses indication stick**>

Now this is taken from John Keith's book and is the death history of transposition. Now you see, here is the number of cases, these are the cases at the age of death and here is the age on the bottom scale. Now you will see that there's a major mortality at the age of under 1 month of age. And you see that 90% were dead at the age of 7 months. Although it's possible to survive over 6 months, few of them do. Therefore if we are to cope with transposition the effort has to be made here.

<**Somerville narrates over chest x-ray, uses indication stick**>

Now, the diagnosis of transposition. You should always think of it if you see a blue infant, a blue neonate. But there is a problem. In the first week of life, both the chest x-ray may look almost normal so you mustn't be put off by this if you think it may be a case of transposition. And you may have to catheterise if you think the baby has this condition because they rapidly do deteriorate and sometimes by the time the chest x-ray obviously […]

<**Somerville narrates over further chest x-ray, uses indication stick**>

[…] shows a case of transposition as in 10 days later as in this case, the baby is gravely ill. Thus the chest x-ray, as the ECG, may be normal in the first week of life and this is important to remember.

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<**Somerville narrates over chest diagram, uses indication stick**>

Now, looking at transposition which when written about I think is made rather complicated, let us look at it in detail a little bit closer. Now here you see a simple diagram showing you what transposition is, and basically it is a defect in plumbing because the aorta is connected to the venous side and the pulmonary artery to the arterial side. So you can quickly see that unless there's some important communications somewhere along the line, life is impossible because you won't get the streams mixed.

And it is on this fact that the basic principle of creating transpositions is based. Now the obvious and best place to have a communication is up here and this is shown in the natural history studies of transposition where those with an A.S.D. seem to survive better than those with other lesions or no lesions. Thus came the principle of creating an atrial septal defect. This initially was done by surgery but now with Bill Rashkind's remarkable discovery […]

<**Somerville narrates over chest x-ray, uses indication stick**>

[…] he has shown us how we can create it by pulling a balloon through the atrial septum. Here is a catheter passed from the femoral vein up into the right atrium, into the left atrium; the balloon is blown up and it is jerked quickly through the atrial septum. And this creates an atrial septal defect and allows mixing.

<**Somerville to camera**>

Now this excellent palliative procedure has saved many lives and it is better than having to open the chest of an ailing neonate. If it is not successful, one may have to do that. If there are other lesions such as large V.S.D., ductus, they may have to be dealt with at the same time.

<**Somerville narrates over previous chest x-ray**>

But this is done at the time of diagnostic catheterisation.

<**Somerville narrates over graph, uses indication stick**>

Now, the early results from the Sick Children's Hospital are shown here. Here's the age, at the time of Rashkind, and here is the number of cases. You see the large bulk falling in the neonatal period. And here, cross-hatched, are the ones that died in spite of this. And you see that quite a number have been saved, in fact the major bulk have been saved, and on analysing the ones that have died, we have found that this is an early part of the series, it is mainly those patients that have additional serious lesions.

<**Somerville to camera**>

So you see that this is an excellent palliative procedure.

Now what about the diagnosis of transposition?

<**Somerville narrates over table, uses indication stick**>

Well, basically I think it should be thought of in any dusky, ill neonate and it may mean that one has to catheterise some babies that do not actually have heart disease. In any baby that is very blue and has a loud murmur, in any baby that has heart failure, 'stiff' lungs and blueness, in any baby that is blue and you look on the chest x-ray and the lungs are full of blood, and later on when you can see the vascular pedicle on the x-ray, any baby that is blue with an abnormal looking vascular pedicle. You must think of the diagnosis of transposition.

<**Somerville to camera**>

But in the neonate, this is a cardiac emergency.

Now, when else does cyanotic heart disease in the infant require help?

<**Somerville narrates over table, uses indication stick**>

Just having cyanotic heart disease does not necessarily require help. We've already dealt with the problem of transposition. If the baby has cyanotic attacks which I'll mention in a moment, if they have extreme polycythaemia – haemoglobin up 18, 19, 20 grams – that's bad because they may lead to thrombotic or embolic incidents, and if they've had one that, in a sense, is an emergency and something should be done. If there's spontaneous closure of a duct which may be keeping the baby with pulmonary [?] alive, that is an emergency. Or if there's spontaneous closure of a man-created shunt, that is an emergency and something must be done.

<**Somerville to camera**>

Now of the other lesions that we should mention, is Fallot's Tetralogy. Fallot's Tetralogy takes death in infancy and it is something that something can be done about and they can ultimately be corrected if saved.

00:30:06:01

<**Somerville narrates over diagram of Fallot's Tetralogy, uses indication stick**>

What is Fallot? Fallot is the combination of high grade pulmonary stenosis and infundibular and valve level with a V.S.D. here. So the blood, instead of passing through the pulmonary artery, tends to go out of the large V.S.D. and the baby is blue.

<**Somerville narrates over angiocardiogram, uses indication stick**>

Now, here is an angiocardiogram which summarises the important anatomy. Here the right ventricle has been opacified with dye and here the dye passes up through the narrow infundibulum, made of muscle, through the narrow valve and out into the pulmonary artery. It also passes across the large ventricular septal defect to opacify the aorta. Now there you see one phase of the cardiac cycle and here you see another. And the striking difference between these two pictures is that the

infundibulum is wider there, so it's not a fixed obstruction. This picture summarises one of the problems of Fallot and this area is very contractile and is capable of shutting spontaneously. If that occurs, no blood will get into the pulmonary artery, or very little, the patient won't be oxygenated and so blue blood will pour out into the aorta. Even more, these cyanotic attacks are a very dangerous part of Fallot's Tetralogy and must be recognised in that they occur in infants of all ages.

<**Someville narrates over table looking at infundibular shut down, uses indication stick**>

They are of sudden onset, they are associated with impaired conscious level, increasing cyanosis, the murmur which you've heard quite easily when the child is quiet may disappear and they get a nasty grey-blue look. They are induced by emotion, effort (sometimes the effort of feeding), colic after a feed or may occur spontaneously. The treatment is sedation, oxygen, now we have Propanolol which may be given in small doses intravenously. But I believe, if you are close to a competent surgeon, that they should have a shunt procedure bypassing the obstructed area.

<**Somerville narrates over table looking at indications for surgical treatment, uses indication stick**>

The indications for surgical treatment in all forms of cyanotic heart disease I believe in the infant are the presence of cyanotic attacks, deep cyanosis, marked polycythaemia (this is a mistake on this slide but you can see what it means); if they're very polycythaemic, as we've already said, they're going to get this – [thrombotic incidents] and it is a terrible thing to see a child with a correctable Fallot who has had a hemiplegia. So I believe we should shunt these patients when they are sick or have any of these features. The actual presence of blueness alone is not an indication if the baby is doing well.

<**Somerville to camera**>

Now, most patients, how are we to recognise which lesion we are dealing with?

<**Somerville narrates over table, uses indication stick**>

The majority of cyanotic conditions are associated with right ventricular hypertrophy. If you see hypertrophy on the electrocardiogram then you should consider such rare things as tricuspid atresia, pulmonary atresia with intact septum and another list of things which don't really cause trouble in infancy. These do, these are important causes of neonatal cyanotic problems and may need urgent help depending on the factors that we have just enumerated. Investigation will make the diagnosis.

<**Somerville narrates over chest x-ray, uses indication stick**>

Here is a patient with pulmonary atresia and intact ventricular septum. Here, the right ventricle is somewhere in here, and the dye has been put in and you see the blind pulmonary atresia. Nothing can get through this and the patient lives on the blood that goes through the left ventricle into the aorta and goes through a duct. And you can see that if this duct closes, as unfortunately it does sometimes, then the baby will be in terrible trouble and this is the emergency we mentioned earlier. That's pulmonary atresia and intact septum.

<**Somerville narrates over a further chest x-ray, uses indication stick**>

And here is tricuspid atresia where the dye passes from the right atrium to the left atrium and to the left ventricle and once again blood gets into the lungs either through a ventricular circuit effect or occasionally through a duct.

<**Somerville to camera**>

Now, I'd like the slide off and I'd like to just summarise what I have said – that we have dealt with the major problems as an introduction in infancy, either the patients get into the 'stiff' lung situation or they may have hypoxic blue problems or they may

have a combination of both. And the point is that once they start to be in trouble, help is needed urgently, accurate diagnosis is the keystone of successful treatment.

Thank you very much.

<**End credits**>