Surgery for Cardiovascular Diseases in the Newborn*

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Dr. Hanlon: We want this to be informal with a good deal of interchange between the audience and the panel participants and among the panel members. Dr. Lev will make introductory remarks which will give us a morphologic basis about which this discussion will rotate.

Dr. Lev: My role is to indicate in a general way, for my colleagues on the panel and for yourselves, the various entities we see at necropsy up to the age of about six months, which is roughly the period my colleagues will discuss. I think it is advantageous to place these entities into four categories. Of course, when I say that, you realize that actually a child may die from many entities up to the age of six months, so it is the whole field of congenital heart disease that we are dealing with. The only entities I am going to mention are the common entities, the things that are seen quite frequently and hence occupy at least a statistical part in our thinking. These can be divided into four groups: first, the group with decreased pulmonary flow; second, the group with increased pulmonary flow; third, the group with the small left ventricle, or small left side of the heart; fourth, complete transposition of the vessels which occupies a place by itself.

When we think of decreased flow to the lungs, we are usually thinking of some complex which includes pulmonary stenosis or atresia. Thus, we may have pure pulmonary stenosis. Of course, in this age group it would be associated with an atrial septal defect anyhow, a fossa ovalis defect. The second one in the decreased pulmonary flow group would be the pulmonary stenosis or pulmonary atresia associated with tricuspid stenosis or atresia. The third is that which is associated with transposition, in other words, a pseudotruncus. Here the aorta overrides or comes from the right ventricle, associated with pulmonary atresia. All these are entities which will occupy the attention of the clinicians and surgeons as to diagnosis and possible treatment.

The second group concerns those with increased pulmonary flow. Here we are dealing with the usual things we see with increased pulmonary flow, except that they are occurring at this peculiar age and constitute in some cases a surgical emergency. Thus, the patent ductus which is seen at
this age, or the ventricular septal defect, or even, although it is rare, an atrial septal defect by itself, may be very important in these early days and produce fatalities. My old friend Ben Gasul used to say that atrial septal defects do not die at this age, that atrial septal defects only cause trouble later. He was wrong. We have seen atrial septal defects get into trouble even in the very early ages. In these entities, of course, we are dealing with left-to-right shunts, either at the ductus level, the ventricular level, or the atrial level. There is increased flow into the lungs with plethora and with that comes pneumonitis; the patients die with a combination of pneumonitis and heart failure.

The third group concerns itself with that very interesting and very common group of small left side of the heart. In the small left side of the heart, we can discern three entities. First, there is premature closure or narrowing of the foramen ovale. That usually kills in the first few days due to the fact that the left side of the heart is very small and the foramen ovale is narrowed or closed. As soon as the blood pours into the lungs with the opening of the pulmonary circulation, the patients die in heart failure. What can be done about that is up to my colleagues to discuss. It is also possible that some of these patients live on to a month. I am not sure of that. I have seen a few cases older than one, two or three days. There are about 35 cases in the literature and thus, it is less common than other entities to be mentioned in this group.

A very common one is what we call hypoplasia of the aortic tract complex, or the hypoplastic left ventricular syndrome of Nadas in which we are dealing with smallness of the left side of the heart from birth associated with aortic stenosis or atresia with or without mitral stenosis or atresia. This is so common that in the statistics of some pathologists, it comprises half the entities they see. In my series of about 1550 cases, I think we have seen about 100 or 150 of them. What to do about that—well, this very common thing will certainly be discussed by the panel.

Next, we have the fetal coarctation complex which is almost as common as the previous entity. In fetal coarctation, we have a long segment of narrow aorta, pre-ductal, with a large ductus, an atrial septal defect in most cases, and with that, a smallness of the left side of the heart and largeness of the right side of the heart. This is a close relative to the hypoplasia of the aorta complex. They are similar from the standpoint of the small left side, except that it is relatively uncommon to have aortic stenosis.

The final entity I wish to discuss and about which our very distinguished panel moderator, Dr. Hanlon, has done so much, has to do with complete transposition. Complete transposition of the ventricles is that condition in which the aorta comes out from the right ventricle and the pulmonary trunk from the left ventricle. We will deal with the simple type only because the complicated types will die anyway; there is little that can be done for them. The simple types, however, in which there is an atrial or ventricular septal defect, patent ductus or any of these in combination, are just as common as the hypoplastic left ventricle syndrome, and between the two, before the age of six months, I would say they make up about 50 to 60 per cent of cases. What to do about this group is indeed a fascinating surgical problem. I think I have covered the subject from the standpoint of the common lesions.

Dr. Hanlon: That is an excellent start. The practical problem the clinician faces, whether the infant is referred by a pediatrician or general practitioner to the surgeon directly or to the pediatric cardiologist is: What is wrong with the child, what can be done about it, and how soon does it have to be done? This is a joint problem of internist, pediatrician and surgeon. Dr. Diehl could give the same general talk about medical management which Dr. Lev has presented about the general pathologic categories. We will ask him to start by
taking as his text a question sent up from the audience. The question describes a newborn, cyanotic infant in congestive failure, treated with digitalis, but the cardiologist felt the failure could not be controlled. Further studies showed transposition, coarctation of infantile type, patent ductus (we presume he means the coarctation is pre-ductal) and an intracardiac shunt. There was severe strain on the left ventricle. Would you operate to relieve the coarctation, and if so, would you obliterate the patent ductus at the same time?

There we have a complex defect, one of those newborns who is in trouble and is categorized by the questioner as being both blue and in congestive failure. He has transposition, coarctation, patent ductus and a ventricular septal defect. How do you approach this particular infant?

Dr. Diesl: Rather than to start with a discussion of this case and certain aspects of this desperately ill newborn, maybe we can make a few general comments as to what causes babies to get into trouble with congenital cardiovascular defects. Babies get into difficulty primarily because of two reasons: (1) heart failure; (2) diminished peripheral oxygen saturation. These two situations may occur singly or they may coexist as factors related to the infant's not doing well.

How do we recognize that the baby is in trouble? First, we must ask ourselves whether or not the baby is in heart failure. The cardinal features of congestive heart failure in infancy are three in number and consist of cardiomegaly, hepatomegaly, and tachypnea. First, a good rule of thumb is that a baby is not in heart failure unless he has radiographic evidence of cardiomegaly. Secondly, in all probability, he will not be in heart failure if the liver is not enlarged. Third, a baby is not in heart failure if he has a normal respiratory rate. The latter I want to stress, since it may be very frustrating to try to count a baby's respiratory rate, particularly if the infant is crying or fussing, is irritable, or if he is hungry. It is very important to count the respiration when the baby is quiet or preferably when asleep. We must also remember that the small baby's respirations are normally more rapid than those of an older infant or child. If the baby does not have tachypnea, you can immediately say he is not in congestive heart failure. However, it hardly needs to be stated that tachypnea does not necessarily mean heart failure. There may be pulmonary problems, compensatory tachypnea because of peripheral oxygen desaturation, metabolic imbalance, and central factors, to mention only a few other causes of tachypnea.

As soon as we recognize the presence of congestive heart failure, we should institute measures to relieve the decompensation. The digitalis preparation of choice is digoxin. If the infant is in only moderate trouble and is not vomiting, the digoxin may be administered orally. However, most babies in congestive heart failure are quite ill and parenteral administration is the route of choice. If the situation is critical, the intravenous route is imperative; otherwise, intramuscular administration is adequate. As a rough rule of thumb, we like to digitalize babies utilizing digoxin parenterally in a dosage of 0.03 mg. per pound of body weight as the total digitalizing dose. With this dosage, we have found that about 75 per cent of babies will be adequately digitalized. The remaining 25 per cent will be either intoxicated or underdigitalized. One-half the total calculated digitalizing dose may be given immediately. In about six hours, after reassessment of the baby's response to the initial dose, a quarter of the total digitalizing dose may be given.

The physician must evaluate what is happening to the heart and respiratory rate and the liver size. Incidentally, in regard to liver size, a good thing to do is to take an ordinary ball point pen, mark the edge of the liver on the baby's abdomen at the initial examination and mark it again each time you come back to check the baby. By so doing, a good reference point is established and resolution of the hepatomegaly
is easily recorded. A strip of lead II on the electrocardiogram is mandatory before each dose of digoxin. Remember, however, that some babies will not show evidence of digitalis effect on the electrocardiogram, yet they may be fully digitalized. On the other hand, there may be signs of digitalis effect on the electrocardiogram or even signs of digitalis toxicity without clinical response and the baby may still be in heart failure.

Finally, provided signs of digitalis toxicity are not manifest clinically or electrocardiographically, the final quarter of digoxin is given 12 to 14 hours after the initial dose. The maintenance dosage of digoxin is approximately one-fourth of the total digitalizing dose given once daily. In the early parts of the management of congestive heart failure, we prefer to give the total maintenance dose in two portions, one-half in the morning and one-half in the evening. If the baby is under-digitalized, the calculated maintenance dose may be administered twice each day until compensation or toxicity is achieved.

Ancillary measures may be used in conjunction with the digitalis. Oxygen seems to be most beneficial in the babies who are having ventilatory problems. It is least helpful in the babies who have diminished pulmonary blood flow with the blood returning to the left side of the heart already fully saturated. The utilization of a lowsalt formula definitely is helpful. A good point to remember is that Similac contains only about 50 per cent of the sodium content that whole cow’s milk formula contains. If a marked reduction of sodium content is desired, the baby may be placed on a formula of Lanolac. This preparation contains only about 1 or 2 mEq. of sodium per liter. If diuretic agents are to be used in conjunction with the Lanolac, one must be very cautious not to get the baby into electrolyte difficulties and salt depletion.

The other reason why babies get into trouble is because of peripheral oxygen desaturation. Having an ear oximeter available is helpful, particularly in babies who are not having obvious syncopal episodes or so-called “tet” spells. The baby usually will not be in trouble because of cyanosis unless the arterial oxygen saturation falls below 65 or 70 per cent. If the peripheral oxygen saturation is below these values, either persistently or intermittently on the basis of right-to-left shunting, the mechanism is present whereby “tet” spells are produced and these babies may very well experience central nervous system complications.

Now, let’s get back to the baby that we were asked about. This baby seems to be in difficulty both because of cyanosis and congestive heart failure. I am sure all of the measures we have just talked about have already been instituted. The combination of defects, that is, transposition of the great vessels, ventricular septal defect, “infantile” coarctation of the aorta, along with a patent ductus arteriosus, is indeed serious. We would all agree that this group of lesions in the same baby makes the cardiopathy incorrectible. However, a palliative surgical procedure may potentially help this infant. He probably is in heart failure primarily because of increased pulmonary blood flow. Banding the pulmonary artery proximal to the patent ductus arteriosus should be of benefit to him. In all probability, in the situation described the ductus arteriosus is carrying a right-to-left shunt, since the coarctation is of the infantile type. Thus, by placing a pulmonary artery band on the main pulmonary artery, there will be an increase in the resistance to flow at that point, tending to increase the left-to-right shunting from the left ventricle to the right ventricle, thereby shunting a greater portion of the arterial blood over to the right side of the heart, where it can go out the transposed aorta. Also, the pulmonary blood flow will be lowered, thereby improving the congestive heart failure. The other problem is whether or not pulmonary artery banding alone would create a sufficient mixing to relieve the severe cyanosis. This raises the question as to whether or not creation of an intratral communication would increase the oxygen-
ation of the blood to the right side of the heart going out to the body and I would surmise that this probably would be beneficial.

**Dr. Hanlon:** Although you are not very sanguine about the prospects, you would think if an operation were done in this child who is cyanotic and in failure, it should be a banding of the pulmonary artery and an attempt to improve mixing at the atrial level by creation of an atrial septal defect. I wonder if Dr. Lev would comment on what he thinks is the reason this child is in trouble. What are your comments on the pathogenesis of the trouble, the mechanism of the bad clinical effect with this combination of defects?

**Dr. Lev:** This is a very seriously ill child. We have here the complete transposition which is the fundamental defect as far as I am concerned. The patent ductus and the coarctation are secondary phenomena. The complete transposition of the vessels means that we have insufficient oxygenation through the coronary arteries, cerebral arteries and renal arteries. If you have enough oxygenated blood going through these arteries, everything is fine. The projected possibility of doing a banding procedure together with a Blalock-Hanlon operation raises the question as to whether that is sufficient if you are not going to close the ductus. If you do a banding and you leave the ductus open, aren't you going to get a flushing of blood into the lungs themselves? What do you think of that?

**Dr. Diehl:** I think this may not be the situation. It depends on the severity of the coarctation and since this is apparently an infantile coarctation of the aorta, the severity of the stenosis may be quite great. It certainly is true that by banding the pulmonary artery you reduce the pulmonary artery pressure distal to the band and with the patent ductus being present, perhaps the shunt will be reversed from a former right-to-left shunt to a left-to-right shunt; if there was formerly a left-to-right shunt, the left-to-right shunt will be augmented. Whether or not the ductus should be closed might be best determined at the time of surgery. I would imagine if you had a large flow left-to-right through the ductus, you would have some difficulty in creating a sufficient gradient across your pulmonary artery band and this might be identified at this time. From a hemodynamic viewpoint, I can see no contraindication to taking care of the ductus at the same time. This perhaps might mean resection of the coarctation as well, which technically may not be feasible through the surgical approach utilized for other procedures which we have discussed.

**Dr. Hanlon:** Dr. Neville, would you care to comment on this? Here you have the pathologist and the pediatrician who have decided the child has a transposition and presumably ought to have the atrial septal defect made or increased in size so the communication will be greater; they also suggest a banding. How would you feel about doing those two things if you were discussing this with the clinician or family?

**Dr. Neville:** I certainly agree with what has been said. Ideally, it would be nice to use a pump oxygenator and with total cardiopulmonary bypass remedy his existing pathology. However, we are all aware that at the present time, the combination of lesions described is totally incorrectible by any surgical maneuver. For palliation, I would band the pulmonary artery and create an interatrial defect in order to improve his oxygenation. I am not certain whether the ductus should be closed. If he has a severe coarctation, he may be deriving some of the blood supply to the lower part of his body from this shunt. In all probability, I would band the pulmonary artery and do a Blalock-Hanlon and leave the coarctation and ductus alone. In other words, first improve his oxygenation; if you band the pulmonary artery in addition to the interatrial shunt, you might increase the left-to-right intracardiac shunt.

**Dr. Hanlon:** If you have the preductal coarctation and a patent ductus and you do anything around that area such as closing the ductus and trying to resect the co-
arctation or, indeed, if you take that combination and do a banding on it—(and we have done bandings on some of these)—the prognosis is extremely poor. Banding is a good operation for a simple ventricular septal defect, but in our hands, and in the hands of others, banding in these complex lesions gives an extremely high mortality. I don’t know whether we have settled this, except to say I would tell the family the prognosis was just about hopeless, but I don’t think that necessarily keeps us from operating. Once everybody understands what’s what, our main objective is not to keep our statistics good, or as Sir Russell Brock once said: “Keep from blotting your copy book.” We don’t mind blotting the copy book; unfortunately, it has a lot of blots on it already, many of them made in attempts to improve very grim situations like this.

From the floor: Do you think that with creation of a septal defect alone, the improvement in oxygenation might permit better myocardial function and control of failure?

Dr. Hanlon: This certainly should be considered. In other words, get in on the right side as quickly as possible, make your defect and get out. In many of the transpositions that are inadequately diagnosed, either because of a simple clinical diagnosis of transposition or a catheterization diagnosis without definitive analysis of all the defects, it may be wise to make the septal defect and get out without trying for definitive correction. Leave the banding out since it is a little questionable whether you may not be defeating your own purpose to some extent with the banding.

Dr. Diehl: I don’t know that I would agree with that completely. I think that myocardial function is certainly dependent upon oxygen saturation, but a major factor is coronary flow. The amount of oxygen saturation increase that can be obtained by a Blalock-Hanlon operation alone probably would not have a significant effect upon myocardial function in this particular baby. I would imagine his coronary flow is probably pretty good and his myocardial failure is not, primarily because of inadequate coronary flow, but more likely because of tremendous volumetric overload on the left ventricle. Because of the transposition, the left ventricle has free access to the pulmonary vascular bed via the unobstructed pulmonary outflow tract. I therefore seriously doubt that the creation of an atrial septal defect alone to augment the intracardiac shunting would be enough to get this baby through the operative procedure. If he did get through it, he probably would not be significantly helped. The problem of severe congestive failure would probably still exist and most likely could best be relieved by a pulmonary artery banding procedure at the same time.

From the floor: It would seem that the essential problem here is the state of the pulmonary circulation. I don’t see how this patient could conceivably, or at least I am not aware, that the patient with preductal coarctation could be helped very much by surgery. It seems to me the case is quite hopeless and I wonder about operating on him at all. Are there cases that have had complete coarctation, I am not talking about mild ones, but complete preductal coarctations that have been helped by surgery?

Dr. Hanlon: I will ask Dr. Lev to comment on this, but the degree and extent of narrowing of the coarctate segment is something that is quite variable. How you can tell preoperatively or even intraoperatively about this is a difficult problem.

Dr. Lev: I think we have to return to the basic premise. The trouble with this child is not that he has a fetal coarctation or a patent ductus. What this child has is complete transposition. The complete transposition, as I stated before, fundamentally means that he has relatively unoxygenated blood going to three vital systems. What has to be done, therefore, is to augment the pushing of oxygenated blood to those vital systems. The secondary phenomenon of a fetal coarctation or patent ductus per se is, of course, of great importance even...
tually, but the emphasis on that is wrong as far as I am concerned. The thing that has to be done, therefore, is to get oxygenated blood to the coronary arteries, to the brain and to the renal arteries; to do that, I would suggest a Blalock-Hanlon in this case. The child is going to die without operative intervention. Of course, if the surgeon decides to do a banding, then the question of the duc tus comes up. If you just do a banding *per se*, what it means is even with the fetal coarctation, you may have a left-to-right shunt at the duc tus level. You are going to flood the lungs again and have the increased flow go into an obstructed left ventricle. You are going to kill the child. That is the reason, as far as I am concerned, that it is either one way or the other. Either you do nothing, that's point 1. If you do something, then it's a Blalock-Hanlon. If you still feel you have to do something more than that, which I am not competent to judge, then you have to do the whole works, a banding procedure, plus a ligation of the duc tus and that's a big job. I would think from a purely objective standpoint, a Blalock-Hanlon would be all that is necessary.

*Dr. Diehl:* Again, I think this baby will not be significantly helped by a Blalock-Hanlon alone. In answer to the question from the audience, the other members of the panel have expressed themselves about this already. I think we have to admit we have a desperately ill baby who is in all probability going to end up in the pathologist's hands. I think the question is whether or not one wants to "take the bat off his shoulder." Do you want to stand up there and do nothing, or do you want to carry out some type of palliative procedure to prolong his life? We are in unanimous agreement that this baby has a surgically incorrectable lesion at the present time.

*Dr. Hanlon:* If I could formulate this as I see it, there are two possibilities, aside from the do nothing. One, you would just increase the size of the atrial septal defect, or you could move on and add a banding. As Dr. Lev indicates, if you go to the banding that leads inevitably to the need to do something about the ductus coarctation; if you band and catch the left ventricle between the coarctation and the band, you put your patient into a difficult situation. When we band with ventricular septal defect, we want to drive some blood over into the left side. We may make such children a little cyanotic at operation, but they generally are not cyanotic after operation, even when they remain improved. We are trying to create in effect a "tetralogy" of a sort when we band for ventricular septal defect. If you do that and have the blood flow obstructed distally by the coarctation, you may have banded your patient into trouble.

Perhaps there are other questions. This is a long time to spend on one very complex and very desperate situation. The justification for it lies in its striking at the root of a major problem: as Dr. Diehl said, are you going to take your bat off your shoulder or are you just going to retire? Are you going to let the baby die, or, hopeless as it seems, are you going to do something about it? This is the essence of the problem.

Then there is the other problem of deciding which baby is in immediate trouble. Which one should you let slide if he is doing reasonably well, and which one should you move in on? It is a question of whether to do nothing, or do something which has a desperate prognosis. I think this is a very difficult area.

*From the floor:* What would you do for a truncus arteriosus?

*Dr. Hanlon:* Would you specify the kinds of truncus for us, Dr. Lev?

*Dr. Lev:* Fundamentally, at this age group, we are concerned with truncus with decreased pulmonary flow and truncus with increased pulmonary flow. According to which one of these you have I am sure the surgeons and clinicians here will decide which kind of procedure is to be done. One could go into the multiple variations, but they are really unimportant.

Pseudotruncus is something else. In pseudotruncus, we have a pulmonary atresia
and the aorta is either overriding or the aorta is coming from the right ventricle. Under those circumstances, you already have a built-in decreased flow which is fundamental. That is the essence of pseudo-truncus, except for those which have large bronchial flow. These large bronchial vessels may possibly give you a normal flow, but not increased pulmonary flow. On the other hand, when you are talking about common truncus, the usual situation is increased pulmonary flow; decreased pulmonary flow, at least in my hands, is quite uncommon.

**Dr. Hanlon:** If you think of a truncus as a big common trunk with two pulmonary arteries coming off, one going to each lung (if this is what the questioner has in mind), Dr. Diehl, what is your approach to this?

**Dr. Diehl:** We must again assume this infant is already in trouble because of high output failure. By definition, a truncus arteriosus always has a ventricular septal defect with it. Dr. Lev has pointed out that the baby probably is in trouble because of high output failure on the basis of increased blood flow. If this baby does not respond to a “tight medical regimen” and does not come out of congestive failure and continues not to thrive, then I think one has little alternative. If you use a surgical approach, the procedure of choice here again is pulmonary artery banding. How the pulmonary artery banding is carried out is directly dependent upon the type of truncus arteriosus—type 1, type 2 or type 3. It depends upon the site of origin of the pulmonary arteries. The pulmonary artery banding procedure is not as satisfactory a procedure in the more complex forms of ventricular septal defect, for example, the ones that are associated with a truncus arteriosus. On the other hand, two of the three babies with truncus arteriosus which we have banded have survived the procedure and have apparently had significant clinical improvement.

**Dr. Hanlon:** You have banded both pulmonary arteries?

**Dr. Diehl:** Yes, both pulmonary arteries. One was a type 1 and the pulmonary artery was banded directly after it took off from the truncus, one had the pulmonary band placed around both vessels together, and in the third case, I believe they were banded as separate blood vessels.

**Dr. Hanlon:** I wonder if Dr. Neville would comment on banding of truncus.

**Dr. Neville:** I think Dr. Lev answered the question quite adequately in his description of truncus—whether the infant has increased or decreased pulmonary blood flow is the crux of the situation. If a truncus exists with decreased pulmonary blood flow, then you are obligated to perform an extracardiac shunt to obtain adequate oxygenation. On the other hand, if a truncus is present with increased pulmonary blood flow, then banding would be the procedure of choice.

**Dr. Hanlon:** What is your experience with attempts in these very young ones at definitive operation for truncus?

**Dr. Neville:** My experience in these small infants with a truncus has been very limited. I remember one child who had decreased pulmonary flow wherein a Blalock type of shunt improved the oxygenation despite the small size of the pulmonary artery. This was performed about seven years ago and at the time I left Cleveland, she was doing very well. I have had no experience with banding both pulmonary arteries to relieve the plethora associated with increased pulmonary flow through the truncus.

**Dr. Hanlon:** I think that definitive operations for truncus, true truncus, which have been done in a restricted number throughout the country have been pretty unsatisfactory and I would think that attempting it in the newborn period would be almost unheard of.

**Dr. Diehl:** I would like to ask the surgeons, as well as Dr. Lev, what is the likelihood of finding a pulmonary artery of adequate size if an anastomotic procedure is going to be required in the form of truncus...
which has diminished pulmonary blood flow?

Dr. Lev: In the case of truncus with diminished pulmonary flow, the rare kind of truncus, the pulmonary arteries that are found are small in size. Of course, there is a whole range of smallness of the pulmonary tree. In pseudotruncus, the arteries are quite small. In pure pulmonary stenosis, the arteries are fair sized. Likewise, in pulmonary and tricuspid stenosis together, the arteries are of fair size. In common truncus, an in-between situation exists, with the vessels in general larger than pseudotruncus, but smaller than in the other conditions mentioned.

Dr. Hanlon: I think anyone who has tried to do an anastomosis in certain of the pseudotruncus patients has been dismayed by working in the hilum and finding these sleazy, thin-walled vessels in which you may laboriously accomplish an anastomosis in a small baby. Then you are uncertain as to whether you have done it well; the child doesn't improve and a week or ten days later perhaps, you have to go back and try the same discouraging procedure on the opposite side. I think this is perhaps the implication of Dr. Diehl's question. It is a very distressing situation. The Blalock type of shunt in the severe tetralogy at a very early age is attended by a high percentage of inadequate results and this is the reason the Potts type of anastomosis, under these circumstances, in the newborn is commonly favored, aside from the fact that it can be done more expeditiously.

I think we should ask Dr. Diehl and others to comment on the problem of putting a child into as good shape as possible and then getting a definitive diagnosis. Then, if the pediatrician refers the patient for operation, the surgeon knows what he is going to operate on. This brings us to the question of definitive diagnostic studies, specifically cardiac catheterization and angiography. I wonder what your indications, contraindications, techniques and so on would be in these very little ones. Do you have any hesitation in seven, six or five pound babies in doing these definitive diagnostic measures?

Dr. Diehl: It is imperative that as much information as possible be obtained in regard to anatomic diagnosis, as well as hemodynamic alterations. This is vital to the surgeon so that he can plan ahead and have a better idea as to what he is going to encounter and what he is going to do once he gets in there. I think the likelihood of the baby's making it through surgery and being benefited by the operative procedure is markedly increased under these circumstances. Simply because a baby has a congenital heart abnormality or some other defect of his cardiovascular system doesn't necessarily mean that the baby has to have cardiac catheterization or angiography. I think this is quite apparent. We do have a rule of thumb that any baby that is in trouble (and we just talked a few moments ago about babies in trouble) should have definitive diagnostic procedures carried out as soon as he is in the best possible shape to tolerate the studies. In other words, once you have relieved the congestive failure or if you have done your best to relieve the congestive failure, this is the time that the diagnostic procedures should be done. If, on the other hand, the baby's course seems to be one of deterioration in spite of what you are doing, you had better work pretty fast toward emergency diagnostic procedures. If one does not follow these ground rules, the occasional baby that has a lesion which can either be cured by operation or can be markedly improved by a palliative procedure will be deprived of this opportunity.

Dr. Hanlon: Do you think you compound the child's problem by cardiac catheterization when he is deteriorating?

Dr. Diehl: I don't believe you do. We don't like to have to put any sick baby through this type procedure. However, accurate diagnosis is imperative. I think there are going to be some babies who will die during the catheterization procedure.

Dr. Hanlon: Have you lost some of these?
Dr. Diehl: Yes, we have.

Dr. Hanlon: Is it a very high percentage?

Dr. Diehl: In desperately ill babies of the age group we are talking about here, we can get them through the catheterization procedure with a risk in the neighborhood of only 2 per cent.

Dr. Hanlon: What sort of anesthesia do you use?

Dr. Diehl: We use a lytic cocktail of chlorpromazine, meperidine and phenergan.

Dr. Hanlon: In the very small ones you give meperidine?

Dr. Diehl: We usually do. If they are desperately ill, we may give them nothing but a sugar nipple.

Dr. Hanlon: I think this is a matter of some importance. I don't know what the general technique is around the country for anesthesia for cardiac catheterization, but we use no general anesthesia for catheterization at all, left, right, young or old. That embraces over 2,000 patients with about 1750 right heart catheterizations in babies as young as three days. They get phenobarbital and also Vistaril through the catheter at the time. Early in our experience, before Dr. Mudd's laboratory was set up, anesthesia was the greatest hazard of the whole procedure. Some of these angiographic studies were done under general anesthesia and there was a forbidding mortality in infants.

Dr. Diehl: I don't think that general anesthesia has a place in this type of diagnostic procedure. If the baby is in trouble, he probably is not going to get out of trouble without some type of surgical procedure. If a surgical procedure is to be done, there are enormous advantages in knowing what the anatomic and physiologic diagnoses are. With the aid of cardiac catheterization and selective cineangiography, the information that is necessary usually can be obtained in a short period of time without undue risk.

Dr. Neville: I do not think we can overemphasize the fact that it is mandatory to know as much about the cardiac pathology as possible before an open heart repair. I well remember the first open heart my former associate, Dr. George Clowes, and I did about ten years ago. The pediatricians asked us to operate upon a severely cyanotic newborn who was going to die unless something could be done to rectify his intracardiac lesion. Without preoperative catheterization which was not done because of the precarious condition of the baby, we perfused the child with our bubble oxygenator. We could not find a cardiac lesion to account for the cyanosis. The oxygenation was striking during the perfusion but the cyanosis returned when the machine was shut off. We did this several times before taking the child completely off bypass. Postmortem examination disclosed agenesis of the pulmonary alveoli with a perfectly normal heart. Admittedly, this was ten years ago, but aside from that, I don't think enough emphasis can be placed on the fact that we should know what the diagnosis is when we operate on these infants.

Dr. Hanlon: I think it ought to be emphasized that catheterization is a safe procedure, if it's done without too much pharmacologic dosage; it ought to be done aggressively at any age so as to give the surgeon something he can rely on. Ordinarily, we don't hesitate, although if the child is in frank failure, I think our laboratory has been hesitant to do it. If after two or three days of what Dr. Diehl has called the "tight medical regimen" with the liver still down and urgent need for information still present, we will catheterize anyway.

Dr. Diehl has referred to the question of whether the baby is in trouble. If, for example, you have the diagnosis of transposition in a baby three or four weeks old, quite cyanotic, hematocrit not very high yet, 52 or something like that, what is your inclination under those circumstances—to recommend creating or enlarging the septal defect or just rocking along to see if the child will get bigger? This is what you are repeatedly being asked. Some people say:
“ultimately this child will have to have a palliative operation. Why not do it now?”
With a good, fat little baby, before it gets into critical difficulty, do you think such a child is “too good” to operate on?

Dr. Diehl: I don’t think we want to kid ourselves into thinking the baby is going to outgrow it. Obviously, this is not going to be the situation. I think what we really have to ask ourselves is: is there a risk in procrastination of several weeks or several months? If the baby does begin to deteriorate, then perhaps the operative risk is going to be greater. Or are we talking about the advantages of waiting for perhaps some solid weight gain, such that the operative risk will be somewhat less at that time. Other factors must be considered. The pulmonary vascular bed in transposition of the great vessels is of great concern to all of us. Perhaps the damage is already done. I am inclined to believe it is probably not already damaged in the early newborn period, but if one waits for several months or until the baby is a year or so of age and is rocking along doing reasonably well, not having central hypoxic episodes and no overt signs of failure, or if it does have heart failure, which is relatively well controlled with digitalis and appropriate measures, then one is concerned whether or not the pulmonary vascular bed is undergoing progressive changes. All of these factors must be weighed and you must try to attach some value to each one of these facets. As a general rule of thumb, once the diagnosis has been made and the baby is in trouble, we will go ahead with diagnostic procedures.

Dr. Lev: I would like to point out as a pathologist what I see in the whole field which needs solution. As far as I am concerned, the problem of increased pulmonary flow is being taken care of at least in a palliative way, and the problem of decreased pulmonary flow is also being handled. The problem of complete transposition in the newborn period is not yet completely under control, at least in Chicago. Of course, we have the Blalock-Hanlon possibility at least to ameliorate the situation. The one thing in my town which is so common and which is not handled at all is the small left ventricle phenomenon. I don’t know how many of them survive— I presume very few, but the point is, in the hypoplastic left ventricle syndrome of Nadas and the fetal coarctation complex, these two, as far as I can see, cannot be handled. I would like to believe that this is a problem that should be solved in the very near future.

Dr. Hanlon: Dr. Neville, will you tell us what you do about this?

Dr. Neville: I am from the same town as Dr. Lev and he has already told you what he sees at necropsy. I have no answer, but the obvious reason the children with hypoplastic left ventricles are seen by Dr. Lev is that we have no remedy for this situation.

Dr. Hanlon: We ought to bring out one entity which Dr. Lev mentioned as a problem in the newborn, that is, total anomalous pulmonary venous return. In correcting this, if you run that blood back into the left atrium and ventricle, perhaps with a small ring at the mitral area, should you make it obligatory that it goes that way, or should you leave the original alternative shunt to the right side open? Although it is my feeling that in the older ones, particularly those done at the age of four or five, you should make a total correction after you have done your shunt, I think in the newborn it may be the better part of valor at times to leave the shunt there after you establish the new communication.

Dr. Neville: I agree that in infants we should establish communication between the common trunk and the left atrium and leave the atrial septal defect open along with the left superior vena cava.

Dr. Hanlon: I am sorry I don’t have any answer for Dr. Lev to the left sided hypoplasia. When we first became interested in transposition, I think in one of our earliest comments we said that transposition was relatively rare. I am glad we put “relatively” in there because it certainly is anything but rare. In neonatal deaths from
cardiac disease, transposition is an overwhelmingly common problem and that's the reason I think palliation is important. What is going to be done later on is a problem that doesn't concern us here. The Senning operation for correction of transposition, which has been done with immediate success in a number of cases, has in the long run been disappointing.

There is a question here about the technique of banding the pulmonary artery. This sort of technical question has not been brought in spontaneously because I don't believe it would be of interest to 95 per cent of the people here. However, I will emphasize one thing; I do not favor a technique for this operation in which the degree of banding is decided by intuition. I feel that physiologic assessment by measurements of pressure above and below the band is essential not only for the patient's sake, but also because you're never going to know, if the child gets past the operation, why he is doing well or otherwise. There is an obligation to quantitate what you have done and to correlate that with the child's progress.

*From the floor:* What rules do you use for measurement?

*Dr. Hanlon:* The first is, can the child tolerate it? You band him down to the point where he has a gradient of 50 or 60 mm. Hg across your band and he has a distal pulmonary artery pressure reduced to 60, 40 or lower. I think that's fine, but sometimes you tighten the band and the distal pulmonary artery pressure goes down only 10 mm. Hg and that is all the child will tolerate. The heart begins to misbehave and you intermittently ease off the band and then cinch it down again; in other words, you do just about what he can stand. The more distal reduction you get, the better off he should be. We sometimes band a little and go back for greater constriction at a second operation. If they won't tolerate banding of the main pulmonary artery, we may constrict one side and go back subsequently and band the other side. Dr. Willman has done this in just a handful of cases.

*From the floor:* What is the best way to watch the indications of pulmonary hypertension in obvious failure? Can you let the youngster just go along?

*Dr. Hanlon:* As I understand the question, if you are going to watch a child with a defect without gross clinical trouble, what is the basis for watching him and what are you watching for?

*From the floor:* Yes, not jumping in and doing surgery right away.

*Dr. Hanlon:* This, I am assuming, is at an age of below two years and you want to wait until he is bigger.

*Dr. Diehl:* I think it is difficult to attach significance on any one particular thing, but of course, the point you are making is whether or not the pulmonary vascular bed potentially is undergoing irreversible changes. The climate directly north of us seems to say that this does occur not infrequently. In Kansas City, however, we have encountered this in only two children with ventricular septal defect whose left-to-right shunt became abolished, a predominant right-to-left shunt was present and therefore, the child was considered at that point to be inoperable. I think this is definitely the exception rather than the rule. When the pulmonary artery and systemic circuit pressures are in a balanced situation as determined by catheterization studies, and the shunt is only from left-to-right, I think you need not be too concerned about irreversibility of the pulmonary vascular changes and the operation is indicated.

Without recatheterization, what parameters can one utilize to determine operability? One can look at the chest x-ray to assess the pulmonary blood flow. What is the size of the left ventricle? What is the overall size of the heart? The electrocardiogram and the vectorcardiogram are very helpful from this viewpoint to know whether or not there is still evidence of left ventricular diastolic overload which again indicates you have a dominant left-to-right shunt. Another is your physical examina-
tion and, of course, with a balanced pressure situation, the second heart sound is going to be narrowly split and the pulmonic component is going to be markedly accentuated. This isn't going to be very helpful to you. On the other hand, if the holosystolic murmur that was formerly present at the left sternal border is no longer holosystolic and has now taken on late systolic attenuation, this may be a tip off that the baby is then developing a lesser degree of left-to-right shunt. The assessment of precordial overactivity, particularly as far as the left ventricle is concerned, is also helpful. Finally, the importance of the apical diastolic rumble which is on the basis of relative mitral stenosis (relative due to increased flow across the mitral valve) must be assessed. If the youngster formerly had a diastolic rumble and it was grade II or III, and as you follow him along at six-month intervals, you find out that this murmur is now diminishing in intensity or is now no longer present, then this is another bit of indirect evidence that the magnitude of the left-to-right shunt is diminishing.

Dr. Hanlon: The group in Chicago, as everyone knows, had a great deal to do with clarifying the picture of the evolution of these lesions. Because Dr. Gasul, whom we mentioned, and his associates were so interested in this evolution, in a sense, I think Dr. Lev can speak as a clinician rather than as a pathologist on this subject.

Dr. Lev: I am not competent to speak as a clinician ever. However, Dr. Gasul had shown, as was discussed by Dr. Diehl, that in general, ventricular septal defect patients up to the age of two years may get into trouble. From two to about 13, they reach a sort of status quo and nothing much happens in most cases. Then, they may get into trouble again and may die in pulmonary hypertension. Of course, he didn't know what happens after the age of 15 because that was out of his field. From what we know of the work of others, there are cases that die in pulmonary hypertension after that, but the question arises, what happens to all the others? Do they all die or would they all die if left alone, or would they get well? We don't know that answer, but I can tell you from the pathologist's standpoint that there are ventricular septal defects that do close. We have that evidence, absolutely, irrevocably; we have had it since 1937. That being the case, and knowing all the clinical evidence that is present that Dr. Diehl discussed and that Ben Gasul saw, it seems to me at least at the present time, one should entertain the possibility that small VSD's will close and that probably large ones will not close. Of course, this still leaves us the various problems of pulmonary hypertension, increased flow, etc., but small VSD's, I think, will close.

Dr. Hanlon: We are following a number of VSD's which are called "Roger's defects" by some people, meaning that the pressures are normal, the shunts are negligible and may be picked up only by the cineangiogram. We are catheterizing them at two, three or five year intervals to see what the evolution of the lesion is. Many have not progressed at all, but we have not absolutely proved that one has closed off in this series. I think on this note, with thanks to the audience and from me to the panel members, we will adjourn.

Summary

Certain cardiovascular entities which may require operative treatment during the first six months of life are here considered by pathologist, pediatrician, and surgeon. Emphasis is placed on lesions not presently correctible; this brings out not only the indications for palliation and the operative procedures available, but also the general approach of the pediatric cardiologist to the infant critically ill with cardiovascular disease. Recognition and treatment of cardiac failure, details of diagnostic studies and choice of operative procedure are debated against a background of the pathophysiology of these lesions.