

Families with congenital heart disease

J L EMERY

Wolfson Unit, Department of Paediatrics, University of Sheffield

SUMMARY Families with congenital heart deformities appear to have more difficulties and stress than those with other deformities. Some problems are intrinsic to the deformity but others are largely related to communication difficulties between the groups doing home support and those in supraregional centres.

Any child born with a deformity or disorder that seems either life or career threatening produces anxieties and distress. Congenital deformity of the heart is now the largest single group of gross congenital deformities, and their families have special problems. Some of the problems that occur in their homes are discussed here; this paper is based on personal experience with bereaved parents over a 40 year period.

It was my practice as a children's pathologist to talk face to face with parents of children on whom I had carried out necropsies. During a total of about 7000 child necropsies, I probably saw the parents of nearly a half. Furthermore, in the hospital we developed a system whereby a specially trained mortuary attendant handled the child and talked with the parents in a humane way. Parents talked to this non-medical woman over a cup of tea in a way that they would never talk to me in my office.

During the past 15 years our interests have become increasingly focused on home deaths rather than hospital deaths and we have been carrying out confidential enquiries at increasing depth on all baby deaths from the age of 2 weeks to 5 years. The home visits started with the cot deaths and when some others were visited as controls it became immediately obvious that people would say things at home that they would not mention in hospital. This particularly applied to personal relationships with their relatives and friends.

An enquiry technique has been developed with Dr Elizabeth Taylor, and the amount of information obtained at conferences in the general practitioner's surgery after these visits is of a completely different order to that obtained previously from interviewing the parents in hospital or by holding a hospital case conference.

Unexpected postneonatal deaths

Congenital heart disease presents to families in two forms; children who present symptoms and are diagnosed in the lying in hospital very soon after birth, and children who are apparently completely normal at birth but present three or four weeks later either as failure to thrive or as unexpected deaths. In any series of children presenting as unexpected deaths in the postneonatal period (cot deaths), there is always a group of children with congenital deformity of the heart and these constitute perhaps about one in 20 of children who so present. The death of these children is unexpected by the parents, although in most cases when one analyses the history from the point of view of gaining weight or symptomatology, some abnormalities in the child's development have been present but not recognised. Before the 'cot death syndrome' became popular, to diagnose an unexpected death as a congenital deformity of the heart was usually a relief to the parents as it constituted an adequate explanation for the death. Now the situation has altered. Parents of virtually every baby who is found unexpectedly dead are now presented with the literature on cot deaths in the hospital receiving rooms or by health visitors, usually before a necropsy has been carried out. When a diagnosis of a congenital deformity of the heart appears, it often disconcerts the parents. There is now a wide public awareness of dramatic surgery on babies' hearts but only successes gain publicity. Parents immediately question themselves as to whether they have missed something and whether, but for some action on their part, the child could have had a life saving operation. Once the diagnosis of congenital heart disease is made, the family is removed from the 'cot death' classification and the whole of the cot death bereavement and

surveillance support network from these people is also removed. There is no other system to take its place. These children have not been under the care of any cardiology unit. As pathologists or paediatricians we can arrange for these parents to have genetic counselling regarding the possibility of further children with congenital heart disease, but this is only of marginal emotional assistance to them. These parents are just as anxious about another child death with their next pregnancy as are classic cot death families and need at least as much support.

Acceptance of a diagnosis

In most children congenital heart disease is suspected in them as newborns and then diagnosed. The situation is similar in general ways to the other major group of congenital deformities, namely children with hydrocephalus and spina bifida, but has differences. Both groups have the shock of loss of expectation and of accepting this—for example, ‘Why me and my child?’—and the insecurity of the future. There is often delayed separation of the baby from the family and friends with a distorted growth pattern of ‘love’ between the family and the child. If the child is kept in hospital with one parent, there is again the distortion in the pattern of love. As the existence of the deformity becomes accepted, there follows the anxiety for the child in older age and financial implications. With the knowledge of increased dependency there is then a growth of emotional attachment to this child with essential compensation and possibly over compensation facets. All these aspects apply to all children with defects, but with the child with the heart deformity there is an added difficulty in acceptance. The child that has spina bifida or hydrocephalus usually has some degree of talipes or other physical deformity that the parent can recognise and so actually sees that the child is deformed, whereas the child with congenital deformity of the heart is often overtly completely normal. The child may be pale or cyanosed but parents are not as familiar with concepts of cyanosis and pallor as physicians and these, in my experience, have very little meaning to them, particularly if it is their first child. Thus the child is to them a normal child but the doctors have been able to recognise that the heart is abnormal.

Doctors often spend a lot of time with parents explaining the anatomy of the heart, they show them ultrasound studies, and x ray studies, and models of the heart. This is very difficult for all but the very rare parent. The parents are not basically interested in anatomy, although they appreciate that the

doctors, and particularly the surgeons, are fascinated by it. Parents frequently tell the doctors that they understand exactly what they are describing when they do not. The only thing parents are interested in is whether the child will grow up as a normal child or not. On this point the physicians and cardiologists are always careful not to make a prognosis of complete normality. The parents are carried along on a wave of need to investigate, which they try to understand, but they are still in a sort of ‘zombie’ state of growing acceptance. During this time the babies are lifted from their home base to supraregional investigatory and treatment centres. In such places modern medical magic takes place and the oracles there come out with a verdict that goes down the familial line—treatment/non-treatment, immediate surgery or not.

Different children need different patterns of care. Some children are sent home to die, most go home under supervision to return for staged surgery, some go home directly, and others go to the units from which they were referred. There are problems regarding all these aspects and it is probably simplest to take them separately.

Special units

LEVELS OF CARE: UNIT, LOCAL HOSPITAL, AND HOME
Highly specialised supraregional units, whether for congenital deformities of the heart or other special disease, create as well as solve problems. They are usually staffed by brilliant people who are enthusiasts, some would call them ‘fanatics’, and gather round them a group of people who they are able to infect with their enthusiasm. This produces a level of concentration and interest in the patient that is not available elsewhere. Therein lies the very value of such units and why they can attain such high technical excellence. Parents recognise this enthusiasm, they see it as a special interest in their own child and the doctors and staff of such institutions become a sort of priesthood to them. It is part and parcel of all such units to know how good they are, to tell each other how good they are. This is all part of the psychology of running any good unit. When a child is returned from such a unit back to its base hospital it is impossible for the parents not to be aware that the child is going back to a lower level of care and attention. No matter how good the original referring unit was, it has not the egocentricity relating to that child’s condition that is present in the special unit. This creates problems of confidence for the parents of the referred child. It is not easy for the nursing and medical staff of the base units to handle the situation, everything in a way mitigates against the parents. The child in the ward is not the

consultant's whole responsibility and it is very difficult for a relative lack of interest not to become apparent. All know that the child had to go to another place for 'special tests' and that the major decisions relating to treatment have been determined from outside. Parents often express to us at home how frustrated they are at what appears to them to be defects in local care, which is in actuality not different from what took place in the specialist unit. They are afraid to say anything or make dissatisfaction known to the local staff because they feel they are completely in their hands.

If the child is sent home rather than to the hospital, the situation is often less difficult but there are frequently great problems in communication. There is a timing situation which is very difficult to overcome. If a child is being sent home a referral letter needs to be sent. This usually goes to the referring unit or referring doctor. Such a letter usually takes four or five days. This produces a delay of about 10 days in getting detailed information about a patient to the people who are looking after the child at home. Some units attempt to convey information to the local people by telephone, some have a special liaison health visitor or social worker staff to do this but this works only in a partial way. Telephone information about a child who is only slightly known to either person is difficult, and to catch and speak to the child's health visitor and family doctor entails a use of time that very few units can afford. If this type of information is relayed to the local hospital liaison staff this only creates another stage in which information is reduced in interest and urgency. The need for a formal letter remains. Currently children arrive home and the local health visitor and family doctor don't know that the baby is home until the parents come for help and information. Without a detailed letter of findings and instructions in hand, both the family doctor and the health visitor are at very great disadvantage. Some units send the parent home with duplicate letters giving information for the health visitor and family doctor. This is by far the best method of handling the situation but some units do not want parents to read what they feel are confidential letters and so will not do this. I am convinced that they are wrong in this. I know of one unit where the ward sister always sent a hand written note by the mother to the health visitor—which worked excellently. We have come across several instances where the child coming home produced quite dangerous levels of difficulty in home care, and some cases where we believe that children have died unnecessarily because of hospital staff not being aware of the precise home situation. There are often tensions and factors at home that parents hide

from hospital staff and hospital social workers, particularly if parents want the child home.

ACCESSIBILITY AND COST OF VISITING

The special units have also another problem in that of ease of accessibility to the family. I well remember when visiting one unit and discussing the family situation with the paediatricians there who said how nice it was to work in a unit where both parents brought the child to the unit on every occasion. This was presented as a sign of excellent communications and care of the child. But seen from the other side the situation was quite different. This unit was so isolated and distant from the child's home that it was virtually impossible to reach it except by car. Special units are very rarely sited at city centres but more usually at some previous isolation unit. One parent cannot take a child in the car by themselves so both parents have to go, and in many instances the father has to take the day off work and lose salary in order to take his wife to a special care unit. Some families can ill afford this. In our community less than half of the parents have cars of their own and so visits necessitate either lengthy journeys involving taxis or persuading some friend or relative to drive the car for them. There are some voluntary organisations that do help in this way but these are not usually automatically laid on.

When the child is admitted to a special unit additional problems of time and expense occur. Overnight accommodation is sometimes provided by the unit but by no means always. It is difficult enough for grandparents and siblings to attend local hospitals when children are admitted but attending distant hospitals is very difficult indeed. We have known grandparents not go to see the children in hospital, but pay for the parents to go, thinking that it is better for the child to see the parents than themselves. We have seen parents go into considerable debt in relation to the cost of travelling to see their children in distant units. It is the middle class parent who is at greatest disadvantage as the extremely poor parent on public assistance usually is able to get assistance and asks for it, whereas the young professional has equal difficulties and often more responsibilities. When the NHS has 'rationalised' services, I have never known the additional cost to the families to be taken into account. We hear stories of families in the United States going into debt, selling their houses and cars because their children need special high cost care but the same does occasionally happen within our own NHS and, with current trends, this could well increase.

SURGERY THAT IS NOT SUCCESSFUL

Surgeons are at great pains always to point out that

surgery is not always successful, and the patient may die if not during the operation, soon afterwards. The parents sign a certificate to permit the operation but while they hear the words that the surgeon says, they have accepted the situation usually in a different way. The surgeon to them is to some extent the hand of the Almighty, they know that he knows that he is not successful every time but they also know that, or at least they think they know, he firmly believes that on this occasion there will be success, and thus although there is a theoretical intellectual acceptance that the child may not survive, it is not an emotional acceptance. Optimism is the name of the game. If the child dies the world falls apart in a way that is greater than if a child dies from an operation related to cancer or something that is known to be lethal in the emotional sense. If the child dies, the surgeon is usually at pains to explain to the parents why the operation failed. It is very difficult for the surgeon not to justify things to himself in what he is saying to the parents, and it is commonplace for the parents to come away from their interview with the physicians and surgeons and make a remark to their friends as to 'how sorry they are for the surgeon' that he was not able to be successful in this instance. Deaths in baby heart surgery are more difficult than in other situations because quite frequently the child has been built up for the operation and at the time of the operation is relatively stable, so that if the child dies the parents have the feeling of having signed the child's death warrant when signing permission for the operation. They know that the child would have survived for a few months at least without the operation, and they know that the operation has eliminated this. They had not seen a child getting more crippled physically or seen their child's head getting larger, or seen the child being less able to walk or pass urine that occur in so many other deformities. The cardiac surgical unit carries an almost unique burden of trust.

One of the saddest comments that comes from parents follows their return to the unit where their child lived and died, whether it be for a heart deformity or leukaemia. They go into the ward where their child was being looked after and see another child in the same bed. They cannot but see that child as a replacement 'love and interest' object for the doctors and nurses, whereas they the parents have no such replacement. If they see the doctors on ward rounds, the death of their own child strikes them much harder with the realisation that the interest and activity of the unit staff has moved on while the parents are still left with their own grief and loss.

With deaths of children with congenital deformities, particularly deaths when they occur in distant

special units after weeks of illness, we have a 'dislocation of grief' situation. This occurs most classically in babies who are born prematurely and die several weeks later. When a child dies far away from the home and takes several weeks to die, the parental and the family grief gets out of step. Quite frequently when things go wrong and death is likely, the extended family accept the inevitability of the death and commence their grieving, while the parents who are visiting in the special unit are affected by the optimism of the nurses and are still communicating with the living child. When the child dies later and may or may not be brought home for funeral, the inlaws, grandparents, etc have gone through the initial shock and anger stage of death and are less tolerant of the parents in their grief. The dislocation of grief and affection is a major problem. It is a particular problem in the provinces when children are referred to very specialist units in London. The nurses of these units have spent a vast amount of time and energy on nursing children who later die, and they too need to go through a bereavement process with each child's death if they are to remain healthy. Good units develop their own support system for such cases. Often there is a dedicated padre who helps the parents in their grief and helps the nursing staff in theirs, and the nursing staff and the parents both attend the child's funeral in the institution where the child dies. The child may even be buried in the special unit area and not go back to its home base. This is fine for the unit but when those parents now go home they are in a completely different situation. They have lost both their child and their immediate home support.

The egocentricity of specialist centre workers can be intriguing! Recently at a meeting I listened to some erudite discussions on the technology in diagnosis and surgery of a particular type of congenital deformity of the heart where one hospital series was compared with the other. Neither series was a total community series and both were 'of unit' results but referred to total survival figures with most of the time at home. The family background and home support mechanisms for the children were in no way comparable. They were in different countries in which primary care systems were different. One country has about a half of the postnatal infant mortality rate and cot death rate than the other. In these two operation series no home care aspects were investigated. It was accepted that the mortality rate in the unit from the country with the lower infant and cot death rate was due to the surgical treatment. The surgeons are in a dilemma: they have almost absolute power in their theatres but almost none in homes in far away

villages and towns. It is easy to ignore what is not seen and uncontrollable.

Children sent home to die

There are clinical fashions of belief regarding terminal care of children, and while this only involves a few of the cardiac deaths we have seen some instances of incredible family distress produced by the current belief that children should die at home. Many families just do not have the home support, the necessary spouse, relatives, and friends to give 24 hour support to a child for more than a couple of days. Very few families have any knowledge or experience of what terminal home nursing entails, they accept the task in innocence and then feel failures when they have difficulty. These are problems more for children with cancer or clinical medical conditions.

Conclusion

There is nothing here new to most health visitors and social workers. The lack of communication between special units and the family doctors and primary care teams is so well known as to be almost a truism. There are, however, some other points sometimes overlooked.

The family of the child with a congenital deformity of the heart has different problems from those with overt congenital deformities, as the deformity is hidden and the parents are more dependent upon the doctors and surgeons than they are with many other lesions.

There is still a need for greater communication between the people in special units and the primary care teams looking after the child at home. In particular the need for written detailed statements about the care of the child to be sent home by hand with the mother and child whenever the child is discharged and consultation with the home team before the child goes home.

It is extremely difficult for supraregional units to appreciate local problems and the home care affects the health and sometimes survival of the child. It is equally difficult for highly specialised regional and supraregional units to do self assessment of the total care of the child. Local and home factors have probably much more effect on long term results than is realised.

Correspondence to Professor J L Emery, The Wolfson Unit, Department of Paediatrics, University of Sheffield, 312 Fulwood Road, Sheffield S10 3BN.