TETRALOGY OF FALLOT—EFFECT ON THE FAMILY

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The progress of surgery on congenital heart defects in the past 20 years has been impressive. A variety of conditions can now be successfully rectified. For some, surgery can be initiated at an earlier age, benefiting the health and development of the young child and reducing the mortality and morbidity. This is so for children with Fallot's tetralogy, who have had increasingly early palliative operations with, in many cases, total correction of the defect at a later date, which restores the patients to apparently normal life, enabling them to take part in almost any physical activity. For a child born today with Fallot's tetralogy the expectation of life is as yet unknown, but compared with a mean life span of only 12 years prior to modern surgical treatment the prospects are encouraging.

The family investigations that were necessitated by our enquiries into the inherited component of congenital heart disease (Boon, Farmer, and Roberts, 1972) allowed us also to observe the social impact on the family of a child born with a severe heart defect. Our aims here, like those of Hewett and Newson (1970) among families with a cerebral palsied child, were to discover the practical problems with which these mothers are faced and to document how the family as a whole and the mother in particular is affected by and overcomes the disaster that has occurred. Thus a baseline would be provided against which in due course the effects would be compared of the very early surgery that is now possible.

This report concerns 100 patients with Fallot's tetralogy and their families living in the Newcastle Hospital Region. Their ages ranged from a few months up to 28 years, so it was possible to elicit the initial response of the parents of the very young on being told of the heart defect; to observe some children before and some after surgery; to enquire into the type of school attended and how much time had been lost from school as a result of the defect; and to observe the resulting occupations of the few adults who had completed their education. At the same time 47 comparison families, ascertained by the presence of a child of the same age, sex, and social status as the *propositus*, were obtained with the help of the family doctors (Table I). We were

thus able to compare the social as well as the genetic aspects of the disease in the affected and normal families.

TABLE I
INDEX PATIENTS AND CONTROL GROUPS BY SEX AND SIZE OF FAMILY

Family	Index Group (100)		Control Group (47)	
	Male	Female	Male	Female
1 child	13	6	6	3
2 children	19	12	4	6
3 "	9	10	7	7
4 "	13	6	6	1
5 "	2	2	1	2
6 "	0	2	0	0
7 "	3	1	2	0
8+ "	1	1	1	1
	60	40	27	20

RESULTS

DIAGNOSIS AND ITS PROBLEMS

Of the 100 patients in the survey, 30 appeared blue at birth and half of these had breathing difficulties for which oxygen was given. In these cases the parents naturally suspected that there was something wrong with the child, though it was not immediately apparent that the cyanosis was due to a heart defect. Indeed the majority of cases of Fallot's tetralogy are not diagnosed during the neonatal period but during the second and third months (Boon *et al.*, 1972) when the presenting symptoms are usually failure to thrive, feeding difficulties, and respiratory infections. In 25 babies the heart murmur was first heard at the maternity hospital where 60 of the cases in the survey were born. In a further 19, the murmur was first heard at the child welfare clinic, and these children were referred to the family doctors who usually explained to the mother that the child had a heart defect and that examination by a cardiologist would be necessary to determine the exact nature of the condition. The remaining 56 patients were brought to the family doctor on account of an infection, usually respiratory, failure to thrive, or anxiety of the mother, and the heart murmur was heard during the routine examination; again the patient was

referred to hospital for a specialist opinion. In this group five mothers were themselves already aware that there was something wrong with the child, and in these families the diagnosis of a serious heart defect was not so traumatic as in those where it was not suspected. The expressions repeatedly used by the majority of parents to describe their feelings when they were told that the child had a heart defect were 'absolutely shattered', 'shocked', 'felt as though there was no hope for the baby'. The mothers appeared to be more deeply affected than the fathers, who said little and tended to keep their feelings to themselves. It was difficult to estimate the extent to which parents worried; many certainly worried a great deal; but 18 who expressed little concern did not understand the gravity of the condition and had difficulty in comprehending the nature of the defect. The publicity given to 'hole in the heart' babies in recent years had drawn the attention of the parents to the occurrence of congenital heart disease, but all felt it was something which could never occur to them except in the four families in which an earlier child had been affected.

Most parents were grateful for the explanations given to them by hospital doctors, but many experienced great difficulty in discussing their problems in a hospital outpatient clinic, and these particularly appreciated the visits in their own homes that were entailed by this survey, for here questions could be asked and answered at more leisure, and many outstanding worries could be alleviated. These worries are found no matter what the intelligence or level of education of the mother. They particularly related to the need for more support and help with feeding problems in the early stages. The part the health visitor plays in providing support requires examination.

When there were other children in the family the addition of a child with a heart defect caused the parents considerable physical, mental, and financial strain. This was particularly so if it was necessary to travel to and from London for operation and follow-up visits. This problem has certainly been alleviated for the parents since operative treatment has become more readily available in the north-east of England, but a hospital visit for those who have to travel with their children up to 50 miles is still a major undertaking.

SOCIAL INTERACTIONS

Eighteen of the index patients were 'only' children, but five mothers of these *propositi* were very young and expected to have further family. It was noticed that these children lacked companionship and had difficulty in making friends, as they were unable to

take part in the normal activities of healthy children. Seven of the 18 were waiting for operation, eight had shown a marked improvement in physical activity after operation and a corresponding increase in socialization, but three derived little benefit (two of the latter were also mentally retarded).

In 58 families the affected child had brothers or sisters; he was usually accepted by them and they helped to look after him, but in 17 of these families there was some jealousy and resentment caused by the amount of attention lavished on the disabled child by his parents. There is a real tendency to 'spoil' these children lest they produce a temper tantrum and so precipitate a cyanotic attack, and it takes great skill on the part of the parents to give them a normal upbringing.

The repercussions in one family where the affected child was also severely mentally retarded were far reaching. The parents did not wish to place the 10 year-old child in an institution but were relieved by her attendance at a day junior training centre. The mother took part-time work to get out of the house for a while each day and the father arranged to do some of his work at home. In this way one of the parents was always at home to look after the affected child during holidays and times of illness, but both were able to benefit from another environment.

The effect of a handicapped child on the recreational activities of the family was considered. It was found that the main restriction to family holidays was a financial one, and in 20 instances the family had never been able to afford to go away—a proportion similar to that in the comparison group in which 11 out of 47 were financially restricted, but a further 15 hesitated to go away until the child's symptoms had been alleviated, and the remaining 65 had day outings, caravan holidays or stayed with relations.

Twenty-nine mothers agreed that their recreation outside the home was severely curtailed when the baby was young and before he received surgical treatment, though a few mothers went out if a grandmother or husband remained at home to look after the child. Restriction was also observed among seven of the comparison families when the baby was young, though to a lesser extent—29% of mothers of *propositi* compared with 15% of the others.

Work outside the home was not usually resumed by the mother until the child had started school or had had surgical treatment, but at the time of the survey 40 of the mothers were working in a part-time capacity and a further six hoped to do so in

due course. Thirty-four per cent of the mothers in the comparison group were working, but they represented a younger age group of mothers. Twenty-one of the younger mothers had several children to look after and were therefore fully occupied at home.

EFFECT ON REPRODUCTION

It might be expected that after the birth of a child with such a serious defect the parents would delay, or refrain from, having further children. The subsequent reproductive experience of the affected and comparison families was therefore examined.

Of the 100 families in the affected series, 53 had no further children after the propositus. In these, one mother had died, four were separated or divorced, and 15 had reached the menopause. Twelve mothers expressed a wish to have another child, and of these two were already pregnant at the time of interview, and six had decided to postpone pregnancy until the affected child was improved in health. Two mothers had been sterilized, and 19 did not want further family. These mothers may be compared with the 20 in the other group in which the matched children were the last in the family at the time of interview. One mother was a widow, three had reached the menopause, nine did not wish to have further children, but seven did want another baby and two of these were already pregnant when visited. A comparison shows that there is no significant difference between the two series in the numbers who do or do not want more children. What is significantly different ($P = 0.03$), however, is that half of those in the affected families who want more children wish to postpone the next pregnancy on account of the affected child.

The numbers of families with four or more children in each group were similar; there were 31 (out of 100) in the study group, of whom eight mothers had been sterilized; in addition, one mother with three children had been sterilized. The other group contained 14 (out of 47) with families of four or more children, but the number of mothers sterilized in this group was only two, a clear reduction in the proportion though the difference is not significant. In those families in which a birth had followed that of the affected child, the interval between the birth of the proband and the next liveborn sibling averaged three years seven months. The corresponding interval between the matched child and his next sibling averaged two years 11 months. The difference of eight months between the means of the two groups is not statistically significant. There was no evidence of a period of infertility or

TABLE II
FAMILY PLANNING AND METHODS OF CONTRACEPTION

Method Used	No. of Mothers
Contraceptive pill	28
Diaphragm	2
IUD	3
Condom (father)	18
Sterilized:	
Tube tie	6
Hysterectomy	3
Vasectomy	1
Rhythm method	13
Coitus interruptus	
No precautions	
Pregnant	4
Menopause	19
Separated or widowed	6

interrupted pregnancies immediately preceding the birth of the affected child.

There is thus a suggestion from these comparisons that there is a slight but not yet significant increase in acceptance of sterilization in affected families, especially in the larger; and that affected families, in which further children are desired, are content to postpone further pregnancy until the health of the propositus has improved, and this is manifested in the slight, but again not yet significant, increase in the birth interval between the propositus and his next sibling.

METHOD OF FAMILY PLANNING

Ninety-nine mothers (one had died) were asked about family planning. Nineteen of the older mothers had already reached the menopause and 10 were reluctant to discuss the subject of family planning, but the younger mothers were less inhibited. Table II shows the different methods of birth control used. Some parents used more than one type of contraceptive and it was interesting to note that four mothers who had previously taken the pill had changed to another method—one IUD, one condom, one coitus interruptus, and one father had had a vasectomy (with no regrets or psychological disturbance). Another father was seriously considering this operation; in these two families there were three and four children respectively. Six mothers of large families had been sterilized, and three had had a hysterectomy for gynaecological reasons.

Advice on family planning had been sought by 31 mothers, who had asked their general practitioner, and in most cases the pill was prescribed. Three mothers went to a family planning clinic, one had sought genetic counselling, and three asked for further advice about family planning during the course of the interview. When the affected child was the first in the family the risk of having further

TABLE III
TYPE OF HOME

	Private	Council	Rented	Works or Special	Total
Fallot's tetralogy	35	46	12	7	100
Controls	21	21	2	3	47
Haemophiliacs*	30	39	7	1	78

* Boon and Roberts (1970)

affected children had usually been discussed with the paediatric cardiologist and reassurance had been given to the parents.

HOUSING

The type of home occupied by the index families is shown in Table III and is compared with the type of accommodation of the control group and of families containing a haemophiliac child (Boon and Roberts, 1970). The three distributions are similar. Of the index families, the 12 living in rented accommodation were the least satisfactorily housed. Five lacked an indoor toilet, two had no bathroom, and three lived in upstairs flats approached by a steep stairway.

Obviously bungalows and ground floor flats provide the most suitable houses for children who are physically handicapped. Stairs create a considerable problem both at home and at school and 23% of the index patients needed help in ascending stairs.

EFFECT ON SCHOOLING

The standard of education achieved by these children has an important effect on the type of work they are capable of doing as adults. It was encouraging to find that the majority were able to

attend normal schools, and only 19 of those over 5 years of age had attended special schools for the whole or part of their school life.

Table IV sets out the operative status of the children, divided into school age groups. There has been a change in the type of palliative operation in recent years, which has made earlier operative treatment possible.

The Brock operation is now the most usual and the most satisfactory of the palliative operations. It can be carried out on quite small infants. The Potts procedure is almost never done, because of the difficulty of following it by a total correction at a later date. Table V shows the marked improvement in schooling after surgery is complete. Only two of 29 children attended special schools after total

TABLE V
NUMBER OF PATIENTS BY OPERATIVE STATE AND TYPE OF SCHOOL ATTENDED

Age Group (yr)	School	Palliative Operation		After Total Correction
		Before	After	
5-7 Infant	Normal Special	1 0	4 3	7 0
Total		4*	7	7
8-11 Junior	Normal Special	0 0	6 6	15 1
Total		0	12	16
12-16 Senior	Normal Special	0 0	8 2	5 1**
Total		0	10	6

* Three not yet at school

** E.S.N.

TABLE IV
NUMBER OF PATIENTS BY OPERATIVE STATE AND SCHOOL AGE GROUP

Age Group	Pre-operative	Palliative Operation*			Successful Total Correction	Post-operative Death	Total
		Potts	Blalock	Brock			
0-4	5	0	0	7	2	2	16
5-7 (Infant school)	4	0	4	3	7	4	22
8-11 (Junior school)	0	0	7	5	16	2	30
12-16 (Secondary modern, comprehensive, grammar)	0	1	6	3	6	0	16
17+	0	7	4	0	5	0	16
Total	9	8	21	18	36	8	100

* Potts operation—anastomosis between the aorta and pulmonary artery

Blalock-Taussig operation—end-to-end or side-to-side anastomosis between a subclavian artery and main branch of the pulmonary artery

Brock operation—closed pulmonary valvotomy through the right ventricle

correction (neither of these on account of the heart defect). Of the 29 children who had had the palliative operation but not total correction, 11 went to special schools. Obviously it is to the advantage of the child educationally if the total correction can be carried out as early as possible. However, the operative risks appear to decrease as the child gets older, and this is another factor among the many to be considered before surgery is contemplated.

Before surgical treatment half the children of school age lost one quarter to one half of their school time, mainly due to respiratory infections, particularly during the winter months; school attendance showed a marked improvement post-operatively.

Another problem which faces the affected child (like other physically handicapped children) is that of transport to and from school. The children attending special schools are provided by the local authorities with a bus or taxi service. The majority of children attending normal schools were able to walk to school and three who had been operated upon cycled in fine weather.

On the whole, the parents were satisfied with the type of school the child was attending; those with reservations were the parents with children at schools for the physically handicapped, some of which were considered to provide an education of lower standard than that of ordinary schools.

PHYSICAL ACTIVITIES AND HOBBIES

Games and physical education were restricted for those children who had been treated palliatively, though a few took part in non-competitive sport if they felt well enough. Following total correction an increasing number of children were able to participate in active exercise.

EARLY ADULTHOOD

Against this rather difficult childhood of interrupted education and restricted activities it was interesting to see the eight patients who had left school at the time of the survey, remembering that these were among the first to have palliative operations performed to improve the circulation.

Three patients in this group were not working at the time of interview. One, a youth of 18, mentally retarded and with a severe kyphosis, had attended a centre for the mentally handicapped for a time after leaving school, but due to his physical condition he eventually had to stay at home. Another, aged 20 with a hemiplegia, remained at home. The third was a girl of 17 who had recently left school. She had also had thyrotoxicosis, an additional complication which had been controlled

before a second operation for the correction of her heart defect in 1967.

Two of this group were married; one, a man of 28, a draughtsman, had a very good job in London; he was the father of a healthy boy. The second was a 21-year-old woman who had an active and healthy child of 18 months. Her pregnancy had been carefully supervised and she had remained well until eight weeks before delivery when she developed swelling of the feet. A 6 lb (2.7 kg) baby was born at 36 weeks, and for a time after the birth the patient was in heart failure which responded to treatment. The activity of the child has created difficulties for the mother, who becomes breathless on exertion and has had to rely a great deal on help and support from her mother and a neighbour.

A youth of 18 had become an apprentice plumber. One girl aged 17 was taking a secretarial course at a technical school, and another aged 22 worked in a clothing factory which she found satisfactory provided she did not become over-tired.

DISCUSSION

The traumatic effect on the parents of giving birth to a child with a gross physical abnormality has been considered at length by Hare, Laurence, Payne, and Rawnsley (1966), D'Arcy (1968), and Walker, Thomas, and Russell (1971). Severe types of congenital heart disease, e.g., transposition of the great arteries, preductal coarctation associated with other cardiac anomalies, and great vessel atresias, present early in the neonatal period but many other types of heart defect are detected later so that in these families the presence of a heart defect may be broken more gently to the parents, the mother frequently being the first person to suspect there is something wrong with her baby (D'Arcy, 1968).

The results of this survey suggest that health visitors could give more support to the mothers in the early part of the infant's life. Routine visits are infrequent, and often different health visitors call at successive visits and are therefore unfamiliar with the particular family problem. There is much to be said for delegating this supportive work in an area to one particular health visitor who would become well acquainted with each family on her list and would come to be treated as a friend and adviser, as often happens with social workers in foster situations. Alternatively, a health visitor attached to a hospital paediatric department might be specially trained to help all families with handicapped children attending the hospital clinic.

It was surprising to find when affected and control families were compared that the birth of a subsequent child was delayed by no more than eight months on average in those families who were anxious to have further children. But a higher proportion of mothers in the affected group were sterilized and there was a desire to postpone pregnancy in a few families. This is in marked contrast to the families with a spina bifida baby where further children are usually avoided (Walker *et al.*, 1971; Timson, 1970). This may perhaps be due to differences in the difficulties of managing the child, in his appearance, or in the risks of recurrence of the condition in subsequent children. The chances of having a second child with spina bifida or an associated neural tube malformation are rather greater than having a second child with any congenital heart defect (Polani and Campbell, 1955; Boon *et al.*, 1972; Stevenson and Davison, 1970). The contraceptive methods employed by the parents were similar to those reported by Walker *et al.* (1971), although our group had more older mothers.

The proportion of families containing other sibs in whom there was evidence of jealousy and resentment of the affected child (17 out of 58) was higher than in the families containing haemophiliacs (Boon and Roberts, 1970).

The schools attended covered the whole range available and the results show a growing tendency for children to be transferred from physically handicapped to normal schools following operative treatment, though it is too soon to assess what effect this will have on educational standards.

There is no doubt that following surgery the psychological outlook of the whole family is changed for the better. The children become more active physically and, due to an improved appetite and circulation, they start to grow more rapidly. The mothers are no longer burdened with a physically handicapped child and are free to lead a more independent life, and it has been shown that many take a part-time job as soon as circumstances permit.

Apart from those children with an additional handicap, mental retardation or hemiparesis, the prospects for future employment should steadily improve with earlier surgery, better education, and less absence from school. Thus the future prospects for children born with tetralogy of Fallot are encouraging. But operative treatment cannot be undertaken lightly when there is still a significant mortality; in the 46 operations (palliative and corrective) carried out on the *propositi* during the survey there were seven deaths.

SUMMARY

A study of the families of 100 children with Fallot's tetralogy shows that though the parents, on account of the later diagnosis, are spared the sudden shock that the birth of a child with a gross physical defect produces, they experience appreciable worry and emotional strain, particularly when there are other children in the family. Their recreational activities are restricted. The effect on subsequent reproduction is not great, though there is a tendency for the mother to postpone having further children after the birth of the affected child and a greater willingness to accept sterilization after three or four children. The changed pattern of surgery brings improvement educationally and socially. The majority of children after final operation attend normal schools and, unless suffering from an additional handicap, all transfer from physically handicapped to normal schools before the age of 12 years.

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