THE FIRST ONE HUNDRED CASES OF TETRALOGY OF FALLOT ADMITTED TO HACETTEPE CHILDREN'S HOSPITAL

Ali Ertuğrul, M.D.*

The total number of cases of tetralogy of Fallot which have been admitted to Hacettepe Children's Hospital, since its opening in 1958, is now more than 230 and the number of patients who have been operated on is a little more than 100 but this paper will be confined to our first 100 cases since some of the patients have only recently been admitted to the hospital and not enough time has elapsed for a proper followup or for evaluation of postoperative results in the case of operation.

In this paper I shall describe the common findings and symptoms in our selected cases and, after that, I shall try to explain how we are evaluating the results of the shunt operation in cases where it was performed.

During a two and a half year period, 104,644 patients were admitted to Hacettepe Children's Hospital and of these 711, or 0.68 per cent, were found to have some form of congenital heart disease. The incidence of the major cyanotic heart diseases in this group of children with congenital heart defects is shown in table 1. It can be seen that tetralogy of Fallot accounts for 64 per cent of the cases in the group as a whole.

Since tetralogy of Fallot accounts for nearly two - thirds of all the cases of major cyanotic heart disease, it seemed worthwhile to examine these cases in order to evaluate what had been accomplished and to investigate what further might be attempted.

The description of this tetralogy was first made by Fallot in 1888. According to his original definition, the tetralogy consists of four simultaneous anomalies : 1. stenosis of the pulmonary artery, 2. intraventricular septal defect, 3. deviation of the aorta to the right, and 4. concentric hypertrophy of the right ventricle.

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TABLE 1

INCIDENCE OF SOME FORMS OF CYANOTIC CONGENITAL HEART DISEASE AT HACETTEPE CHILDREN'S HOSPITAL

Defect	Number of Cases
Tetralogy	134
Atrio - Ventricularis Communis	14
Truncus Arteriosus	14
Total A.V.R.	12
Tricuspid Atresia	8
Single Ventricle	7
Hypoplastic Left Ventricle	7
Transposition of Great Vessels	6
Ebstein's Anomaly	6
Total	208

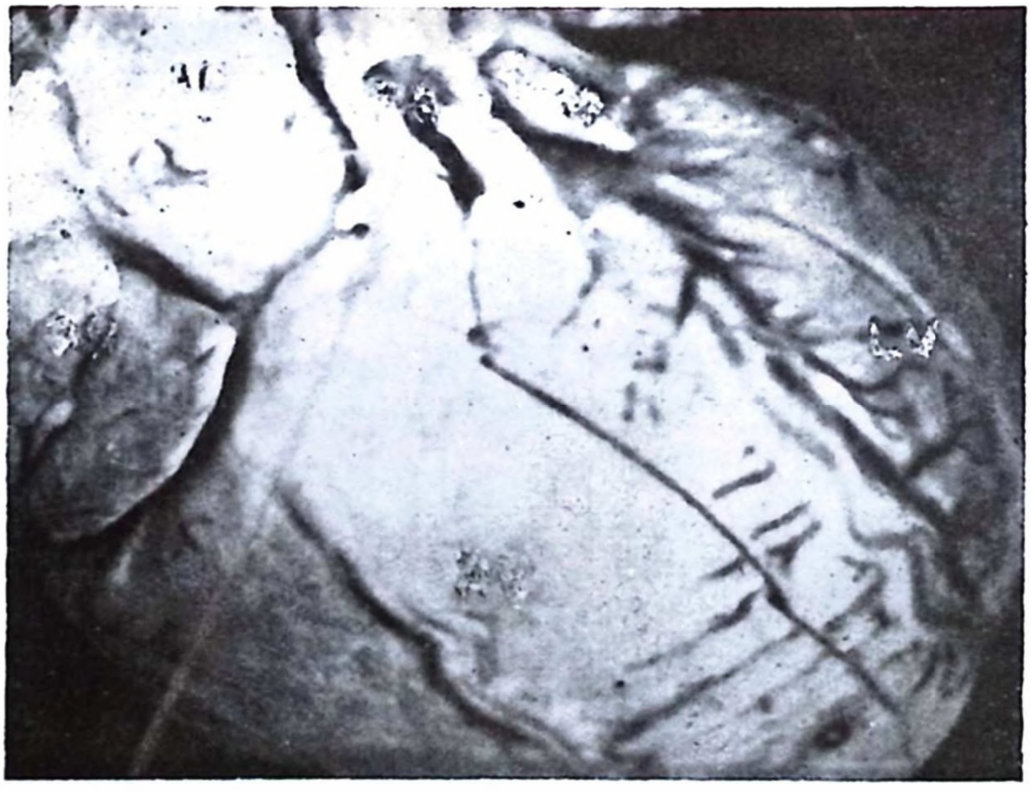

Fig. 1. The tetralogy of Fallot as seen in an autopsy specimen.

Figure 1 presents the picture of tetralogy of Fallot as it is seen in an autopsy specimen. Notice the enlarged aorta and the right-sided aortic arch, narrow main pulmonary artery and its branches, enlargement of the right atrium, and, especially, enlargement of the right ventricle. The dividing line between the two ventricles is identified by the anterior descending branch of the left coronary artery. The left ventricle is displaced a little bit upward.

Stenosis is infundibular in the vast majority of instances. Perhaps pure valvular stenosis is not associated with tetralogy of Fallot as often as is sometimes thought. Our angiocardiographic studies clearly show that the infundibular chamber is a long and narrow channel which contracts during systole and fails to relax

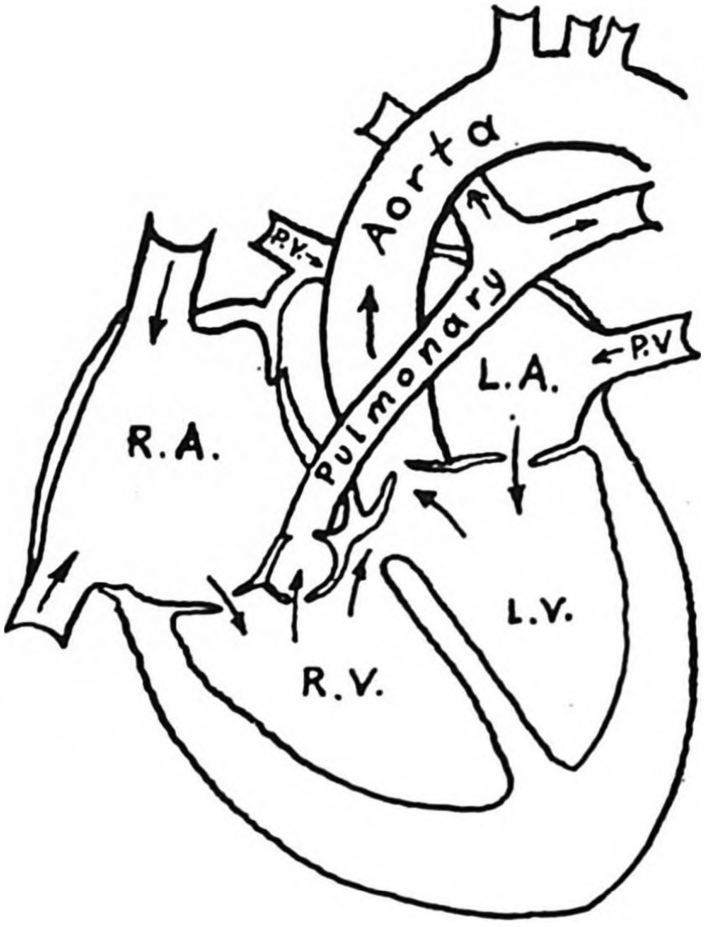

Fig. 2. Diagram showing defects found in tetralogy.

completely during diastole. Of course ventricular septal defects of various sizes are always present in the membranous part of the septum. The degree of overriding of the aorta may be difficult to determine; it is even difficult to determine at postmortem examination. As a matter of fact, because of the high position of the ventricular septal defect, the aorta communicates with both ventricles. In most

cases the thickness of the right ventricle is about equal to the left ventricle and sometimes it is thicker.

Figure 2 is a diagram of the tetralogy. In the aorta, of course, the blood is mixed. Hemodynamic and physiologic changes in tetralogy of Fallot can be explained with this diagram. From the hemodynamic point of view, the essential factor is that the blood flows from the right ventricle to the aorta due to the increased resistance of the pulmonary system which makes it more difficult for the blood to enter the pulmonary artery than to go through the ventricular septal defect and into the aorta. This results in arterial unsaturation, a decrease in pulmonary blood flow and an increase in systemic flow. These patients have identical systolic pressure levels in the right ventricle and the systemic arteries. The size of the right to left shunt through the ventricular septal defect depends on three factors : 1. the resistance in the blood circulation, that is, resistance in the pulmonary system and systemic circulation, 2. the size of the ventricular septal defect, and 3. the degree of overriding of the aorta. This third factor can be explained on the basis of the size of the ventricular septal defect. Due to decreased pulmonary circulation and increased systemic unsaturation these patients have marked limitation of exertion. During exercise the tissue oxygen requirement is increased but the oxygen saturation remains the same, possibly less than at rest in these patients, and exertional dyspnea develops. The presence of the ventricular septal defect is beneficial to the patient. Whenever the demands of the body for oxygen are increased, or when the right ventricle is working hard, the ventricular septal defect acts as an «escape valve» and therefore right ventricular failure does not develop. In pulmonary pure valvular stenosis without a ventricular septal defect and with high systolic pressure, there is no such escape and consequently right ventricular failure may develop. This is why even very severe cases of tetralogy of Fallot, with markedly limited activity, never develop right ventricular failure.

The most extreme degree of pulmonary stenosis is atresia of the pulmonary artery, which is called either pseudotruncus or truncus arteriosus. It may be impossible on the basis of clinical examination alone to determine whether or not there is any blood flowing through the pulmonary orifice. In these cases the clinical picture is entirely dependent on the size of the pulmonary flow and it is very similar to the picture of tetralogy. Since we have actually done only a few angiocardiograms perhaps some of our patients really have pseu-

dotruncus. Pseudotruncus cases get their pulmonary blood flow from bronchial, intercostal and internal mammary arteries and some other collateral circulation, and in most cases there is a patent ductus which carries blood from the aorta to the pulmonary artery. Since there is no clinical difference between pseudotruncus and tetralogy of Fallot we have classified them as tetralogy if angiocardiography was not done. From a practical point of view it is important to know whether the pulmonary artery is present or not in order to know if it is possible to perform a shunt operation. If there is any such doubt, selective angiocardiography should be performed.

As a compensation for anoxia these patients develop polycythemia and hyperhemoglobinemia. If the hemoglobin is more than 15 Gm./100 ml. this is considered by us to be hyperhemoglobinemia and if the red cell count is more than five million we consider this to be polycythemia. Some of the patients have as many as eight million red cells and 24 Gm./100 ml. of hemoglobin. If the stenosis is so severe and collateral circulation is not sufficiently developed, most of the children die during the first year of life - mainly from hypoxia of the brain. Later on, death is most often due to cerebral abscess or, especially in Turkey, to pulmonary or gastrointestinal infections.

In table 2 we see the common findings in our 100 selected cases of tetralogy.

TABLE 2

COMMON FINDINGS IN 100 CASES OF TETRALOGY OF FALLOT

Symptom	Number of Cases	Per Cent	Finding	Number of Cases	Per Cent
Cyanosis	100	100	Murmur	100	100
Squatting*	74	91	Clubbing*	1 yr. 10 1 yr. 90	10 91
Behavior Disturbances*	92	84	Growth Retardation	100	84
Dyspnea	100	73			
Palpitation	100	47	Thrill	100	25

* Percentages, in these instances, are based on the number of children old enough to display the symptom or finding in question, rather than on the full group of 100 children.

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Cyanosis in tetralogy is always due to arterial oxygen unsaturation and increased amounts of reduced hemoglobin. These patients turn deep blue when they cry because this exertion increases the

amount of the right-to-left shunt. These patients never become pink in an oxygen tent nor will their blood samples show a rise to normal oxygen levels after the inhalation of 100 per cent oxygen.

Cyanosis is visible in the finger tips, toes, ear lobes, nose and lips and, in boys, the glans penis. But cyanosis is most pronounced in the external canal and the ear drum, and in the mucous membranes of the mouth and throat. Some patients have redness of the lips and cheeks in a state of cyanosis and their fingernails may even be pink too. This type of patient has less oxygen unsaturation in his systemic circulation than obviously cyanotic patients. In this latter group of patients, the external canal and the ear drums and the mucous membrane of the mouth and throat show a more bluish color than normal, which is useful in diagnosis.

Only 74 patients out of the 100 squatted but the percentage is recorded as 91. This is because 16 patients were under two years of age and had not yet learned to squat, we therefore discounted them in evaluating this particular symptom. Ninety-one per cent of the 74 patients over two years of age exhibited squatting. When such patients are tired they are not comfortable when they sit down.

Very few patients with tetralogy of Fallot fail to exhibit squatting. This symptom was first described by Helen Taussig in cyanotic congenital heart disease and in the tetralogy of Fallot.[1] The first appearance of squatting is usually after two years of age when the child begins to walk, which is later than normal in these children. Most of our patients begin to walk between two and three years of age but recently I saw a patient who is just beginning to walk at five years of age. Dr. Nadas says that squatting begins to disappear at eight to ten years of age due to social pressure.[2] In our 25 patients who were nine years of age or older only three were not squatting and 22 were still squatting even though some were 15 to 16 years of age. Perhaps we have less social pressure against this behavior in Turkey due to the fact that squatting is a common social practice among Turkish village people. We believe this observation supports Nadas' theory. Sudden decreased oxygen saturation due to exercise can be returned to usual levels for these patients more rapidly in the squatting position than in a standing or sitting position. Possibly this is accomplished by : 1. increasing venous return, 2. augmenting peripheral arterial resistance and, by means of this, decreasing the amount of the right-left shunt, and 3. creating pressure on the femoral arteries thereby decreasing the blood supply to the legs and increasing

the blood supply to the brain. In this manner more adequate oxygenation of the brain is possible.

Behavior patterns were noted in detail in 92 cases and among these 92 patients, 84 per cent had some behavior disturbances. In the other eight patients there was no mention of behavior disturbances due to their very young age. The disturbed child is generally very irritable, cries excessively, seems unfriendly, rarely smiles and is afraid of people and generally distrustful.

Dyspnea on exertion is typical for patients who have arterial unsaturation in the systemic circulation. The demands of the tissues for oxygen increase with exercise and, in tetralogy, this demand is impossible to meet. Thus dyspnea and hyperpnea develop. This dyspnea and hyperpnea may be produced by the action of some chemoreceptors in the blood. Seventy - three per cent of our 100 patients complained of dyspnea.

Palpitation can probably also be explained on the same physiological basis as dyspnea. In an attempt to supply the greater oxygen needs of the tissues, the heart works harder during exertion than at rest. Only 47 per cent of our patients complained of palpitation, however, a number of them were too young to make such a complaint.

As to murmur, all of our patients had some kind of systolic murmur over the precordium. Some of them had a grade III murmur, a few had a grade I systolic murmur, and most of our cases had grade II. We have not seen any cases with grade IV systolic murmurs. The classification of murmurs ranges from grade I, representing the faintest murmurs, to grade IV, the harshest.

We have seen no patients with diastolic murmurs, not even in the case of one patient who had no pulmonary artery (observed at operation when it was found that the shunt could not be performed). This patient simply had an enlarged collateral circulation, mainly enlarged bronchial arteries, but even he had no diastolic murmur.

Clubbing is another very common finding in these children. Under one year of age we had only one patient with clubbing of the fingers and toes but over this age we had 90 children, 91 per cent with clubbing.

Eighty-four per cent of our patients had some kind of growth retardation as can be seen from figures 3 and 4 which show the retardation of weight and height in both boys and girls.

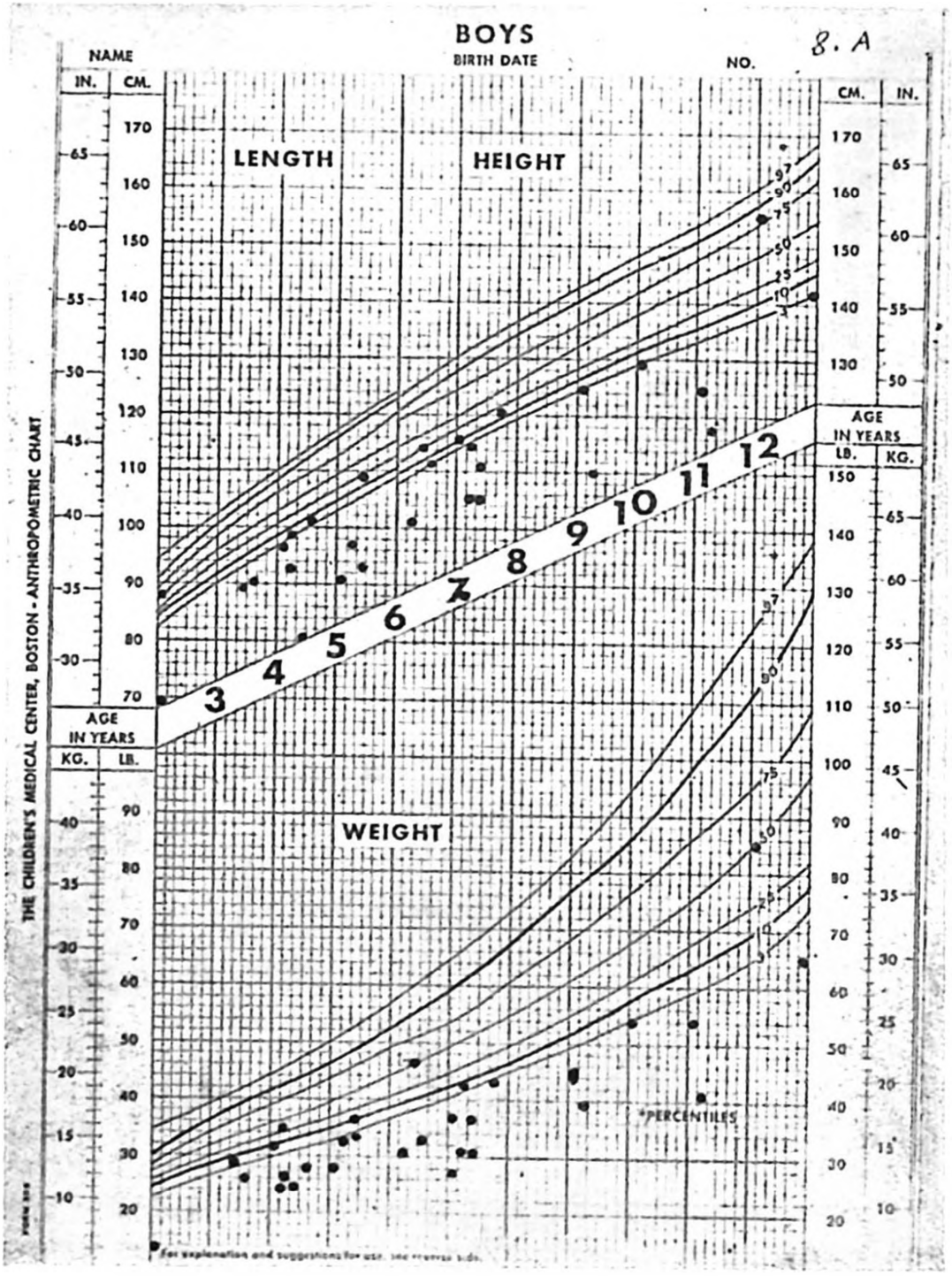

Fig. 3. Growth and development charts showing weight and height retardation in 31 Turkish boys with tetralogy.

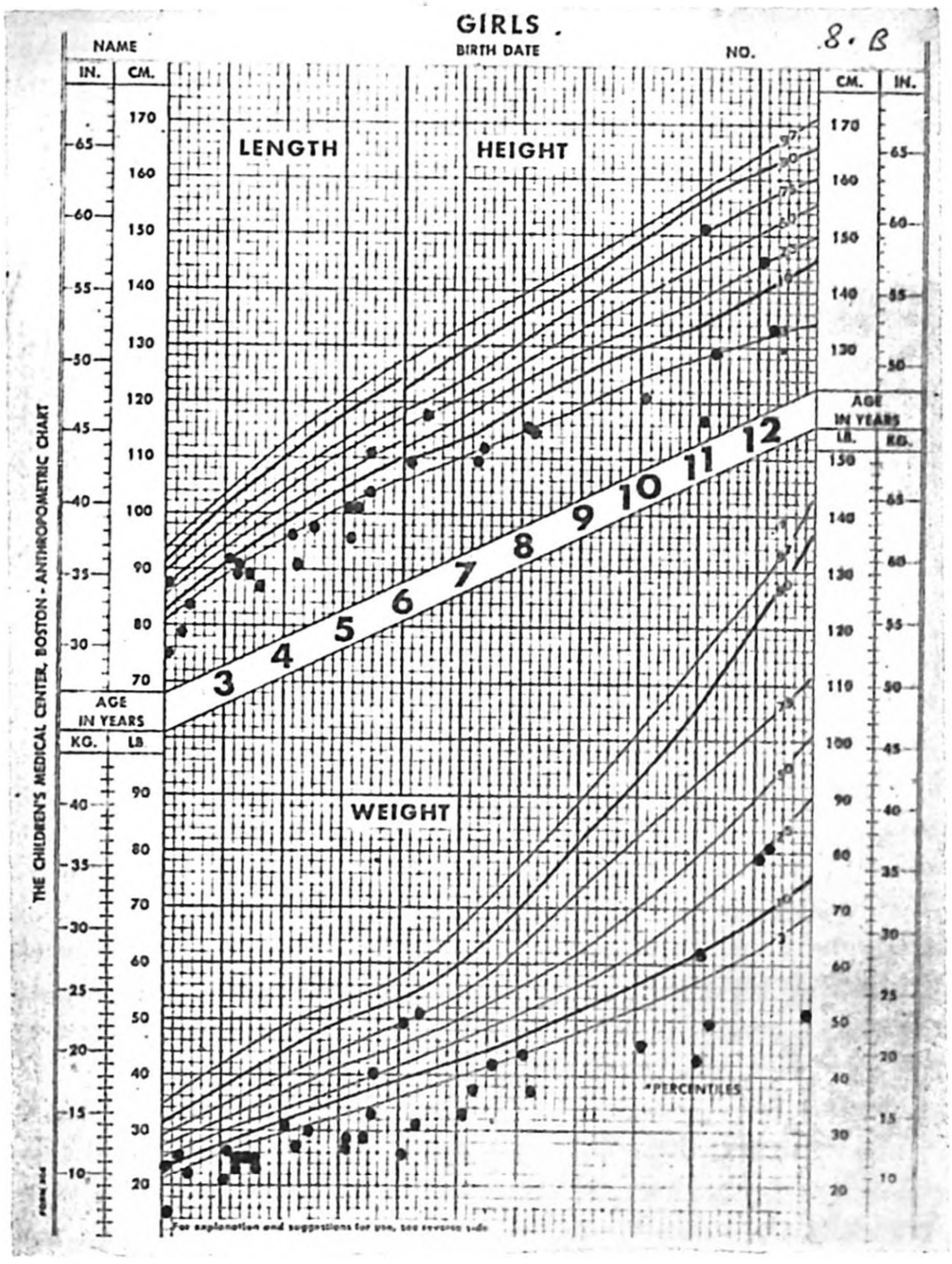

Fig. 4. Growth and development charts showing weight and height retardation in 35 Turkish girls with tetralogy.

Deformity of the chest seems to be a less common finding. Under two years of age we had sixteen babies, none of them with deformities of the chest. Over two years of age, in 84 patients, we found 30 per cent with deformities of the chest, most of them being pigeon - breasted. Some of them had bulging of the precordium. This raises the question of whether age might be the main factor in the development of chest deformities but the number of patients is too small to prove anything conclusive.

The thrill that we have felt was always systolic and as only one out of four patients had a thrill this was not a very common finding.

Blood studies revealed that most of the patients had hemoconcentration. Eighty-eight per cent of the patients had more than 15 Gm./100 ml. of hemoglobin, the red count being more than 5 million in 66 per cent and the hematocrit more than 45 in 86 per cent. Since iron deficiency anemia is quite common in Turkey this might explain why some patients did not exhibit hemoconcentration.

In Figure 3 we see growth and development charts showing the weight and height of 31 boys. This chart was devised by the Children's Medical Center in Boston. It was designed for American children but the observations which we have made at Hacettepe Children's Hospital indicate that in Turkey middle class children under three years of age follow this same pattern quite closely. However, a large proportion of the Turkish population lives in villages and is undernourished. Measurements of village children are lower than the standards indicated on the Boston chart. As can be seen from the plotting of the weights on this chart, only three boys are beyond the tenth percentile and none is beyond the 50 th percentile. There are five between the third and tenth percentiles and the remaining 23 children are below the third percentile. For example, the weight of one boy at age 11 is equal to the 50 th percentile for a 5 year old. The weight of most of these children corresponds to the third percentile of children three to five years younger than they are.

Retardation in height appears to be less pronounced. Five boys are beyond the tenth percentile and between the third and tenth percentiles there are seven. If we take an individual and look at his weight and height together it is again clearly apparent that retardation in weight is greater than in height. There are only a few cases in which height retardation is greater than weight.

Figure 4 shows height and weight retardation in girls. It includes 35 girls from two to thirteen years of age. There are six girls whose weight is beyond the tenth percentile and twelve beyond the third percentile. This indicates that girls' retardation in weight is less than boys'. Also in height the girls are ahead of the boys. It is clearly seen from these two charts that there is retardation of height and weight. But, we believe that tetralogy of Fallot is not the only factor

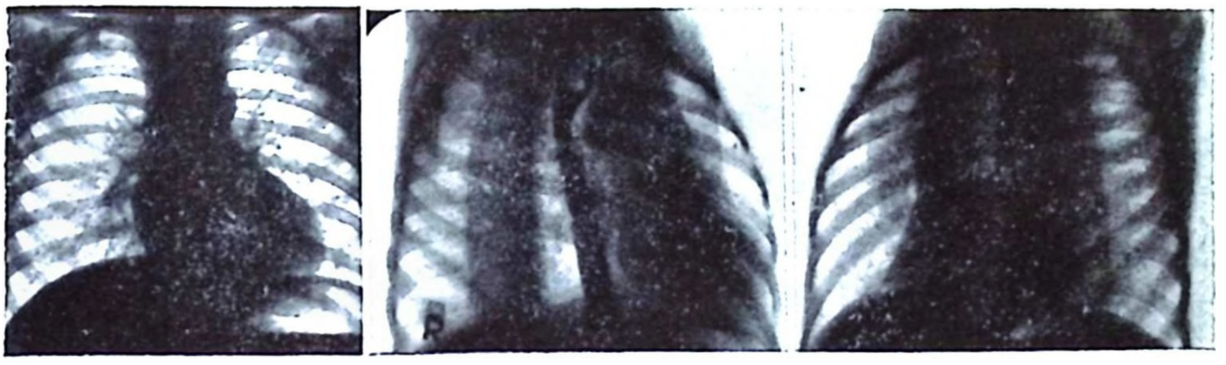

Fig. 5. Three roentgenograms of the same child taken in anterior posterior (left), right anterior oblique (middle), and left anterior oblique (right) with barium swallow.

in this retardation. Some of these patients come from poor sections of town or from the villages where they are undernourished anyway and, probably, this is a contributing factor.

Roentgenography

The roentgen-ray appearance of tetralogies is very typical. Most of our 100 cases can be characterized as follows : 1. enlarged right ventricle with upturned apex *(coeur en sabot),* 2. decreased, or lack of, prominence of the pulmonary artery with sparse vascularity of the hilar region and the periphery of the lungs, and 3. enlargement of the right auricle and an enlarged aorta in many cases. Of course there are many individual variations from this description.

Figure 5 is a roentgenogram of a three year old girl taken with a barium swallow. The heart as a whole is probably within normal limits or, possibly, a little smaller than normal. There is a concavity where the main pulmonary artery should be, the apex is turned up and the aortic arch is on the left side. Also there are decreased shadows of the pulmonary artery and the vascularity of the hilar region and the periphery of the lungs is sparse. The reticular type of vascularity

in the lungs could be due to collapsed circulation. As you can see in the middle film, taken in the right anterior oblique position, there is no left atrial enlargement and the border of the heart is closely related to the anterior wall of the chest. In the third film, which is of the same patient in a left anterior oblique position, the heart shadow is

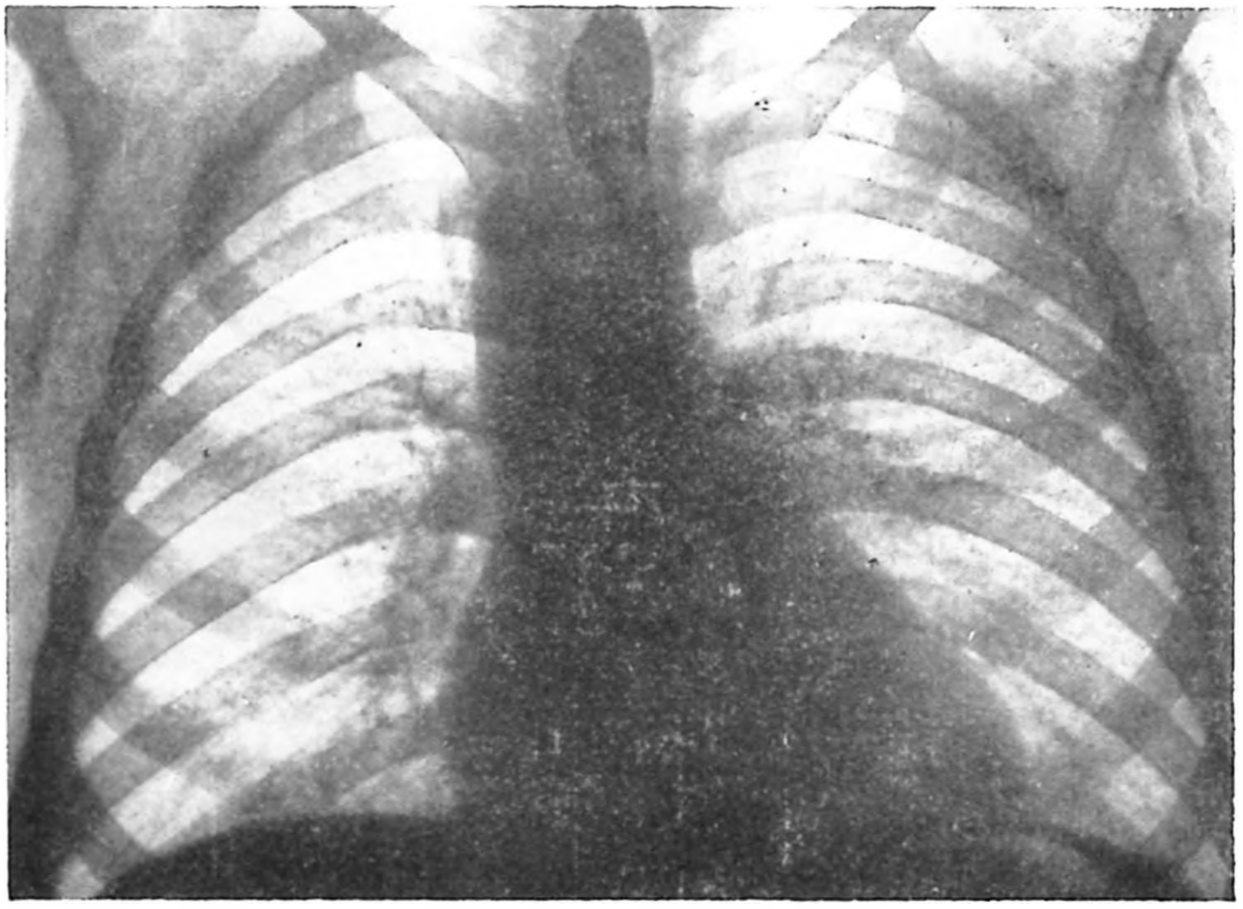

Fig. 6. Another roentgenogram with barium swallow showing aortic arch on the right side.

pushed back and overlaps the shadow of the vertebral column. We think this is not hypertrophy of the left ventricle because the enlargement is in an elevated location, not in the expected lower position of left ventricular hypertrophy, and therefore it is most likely due to right ventricular hypertrophy. This is called pseudo-left ventricular hypertrophy.

In figure 6 we have a roentgenogram in which you see a heart; probably within normal limits or perhaps a little bit larger than normal, with decreased vascularity of the lungs. This vascularity is definitely of the reticular or sparse type. Without barium swallow

it would have been difficult to decide whether the aortic arch was on the right or left in this film but with barium swallow, it can be clearly seen that the aortic arch is on the right side. There is an aortic indentation on the right side of the barium-filled esophagus and the descending part of the thoracic aorta can be seen to the right of the vertebral column. This leads us to believe that in order to say anything about the position of the aorta, a barium swallow film must be taken and that the x-ray employed must be of high penetrating quality.

Angiocardiography

The two films in figure 7 belong to the same patient, a two year old girl. The angiocardiograms were taken simultaneously, five pictures in one second, anterior, posterior and lateral. These films were taken during diastole and the tip of the Lehman catheter is in the infundibulum of the right ventricle. The dye is mainly in the right ventricle and it can be seen passing through the ventricular septal defect into the left ventricle simultaneously. There is also a hypertrophic crista supraventricularis and notice the trabecular hypertrophy in the right ventricle. The main pulmonary artery and the infundibulum are fairly small. If the film had been taken during systole it would have been possible to see that the infundibulum is narrower than it appears to be in figure 7. The pulmonary artery and the aorta fill simultaneously with dye and it is obvious that the pulmonary artery is much smaller than the aorta, about one quarter the aortic size. The pulmonary artery in normal persons is, as everybody knows, at least as large as the aorta or one to two millimeters larger than the aorta. The crista supraventricularis is very thick and it narrows the infundibular channel. The carotid arteries, the aortic arch and the descending part of the thoracic aorta can be seen.

Electrocardiographic Changes in Tetralogy of Fallot

First of all I would like to emphasize that we have ever seen a normal EKG in cases of tetralogy of Fallot. The EKG in these cases is characterized mainly by right axis deviation, vertical heart position, right ventricular hypertrophy, P congenitale, and clockwise rotation.

This EKG shows the typical changes in tetralogy. Here there is right axis deviation, vertical heart position, right ventricular hypertrophy and clockwise rotation. It should be noted that although

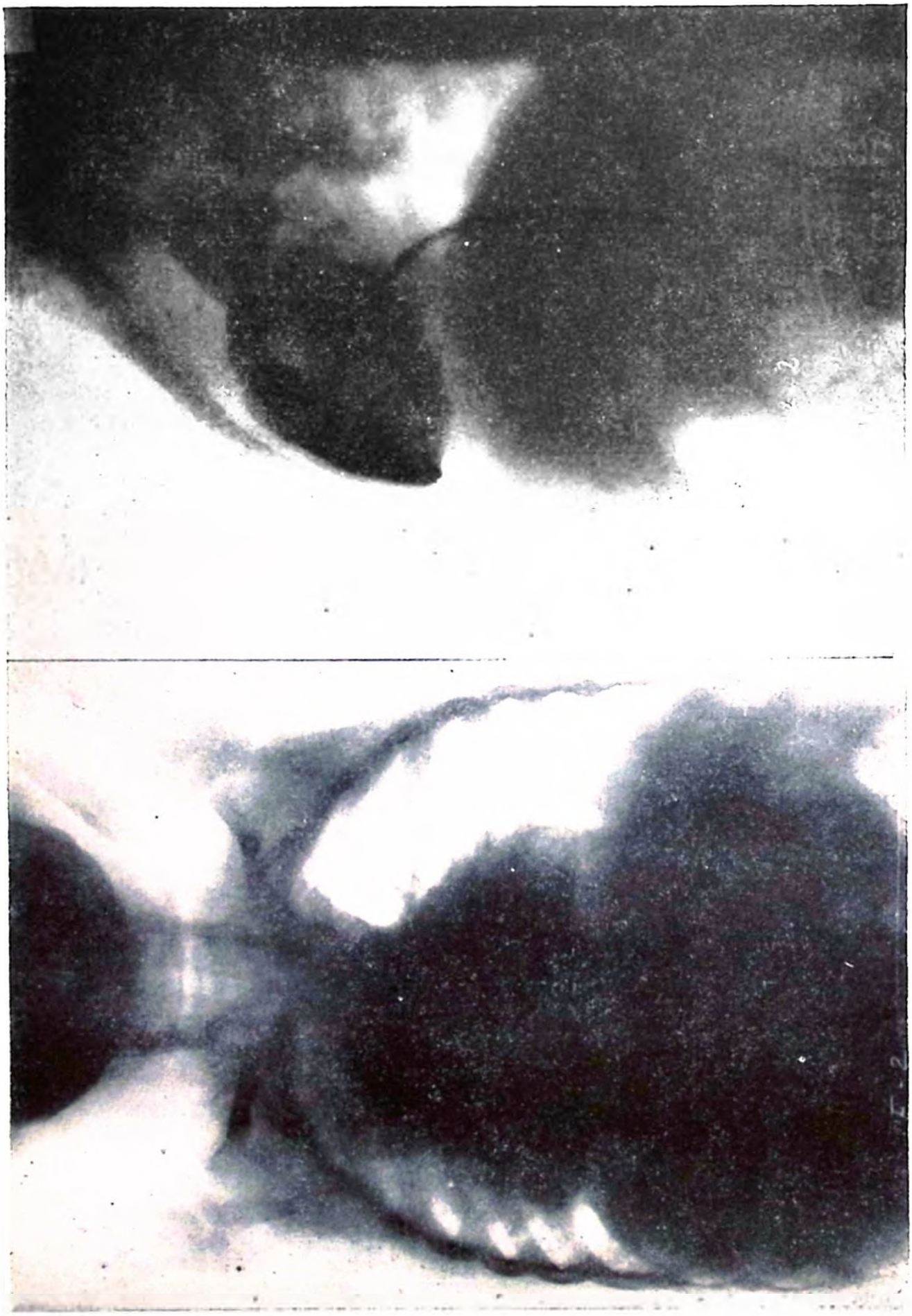

Fig. 7. Simultaneous selective angiocardiograms of patient with tetralogy. Right hand film is lateral, left is anterior - posterior.

abnormally peaked and tall P waves and vertical heart position are typical they are not necessarily always associated with tetralogy.

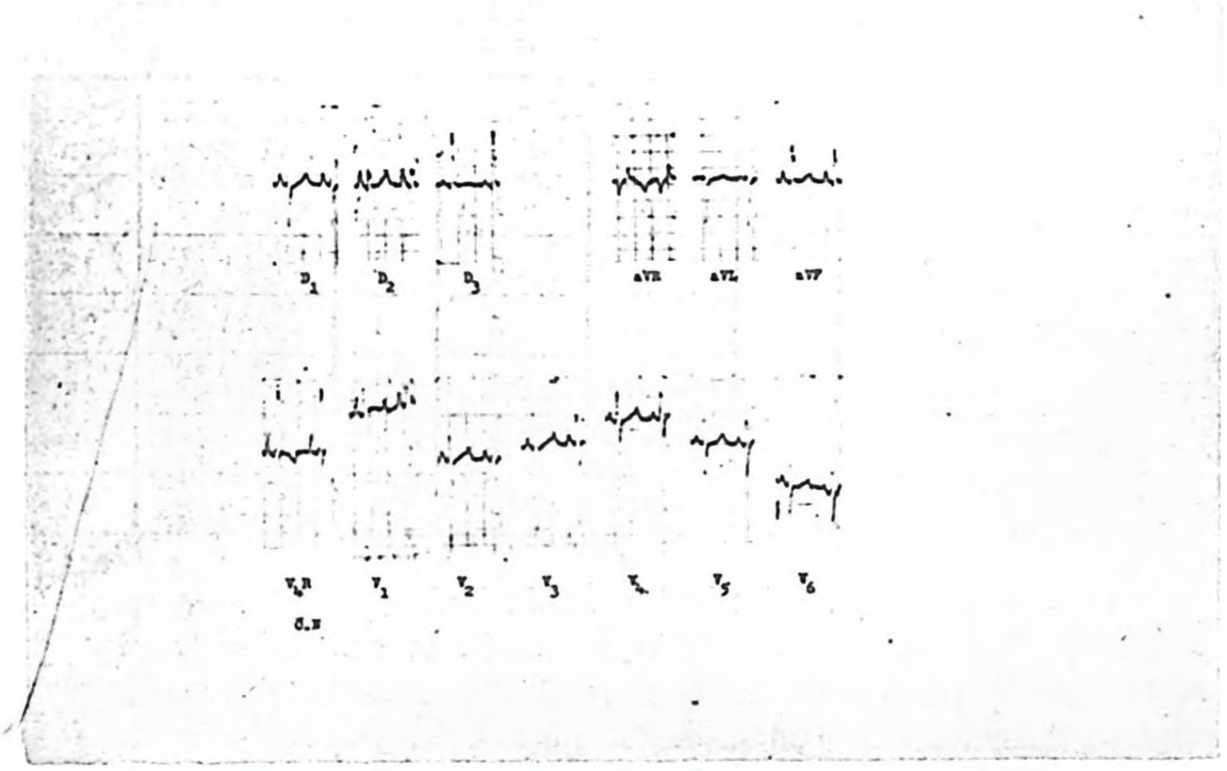

Fig. 8. Typical electrocardiographic changes in a case of tetralogy.

In figure 9 we see that in some cases the T waves may be negative in the left precordial leads as you will note here in V5 and V6.

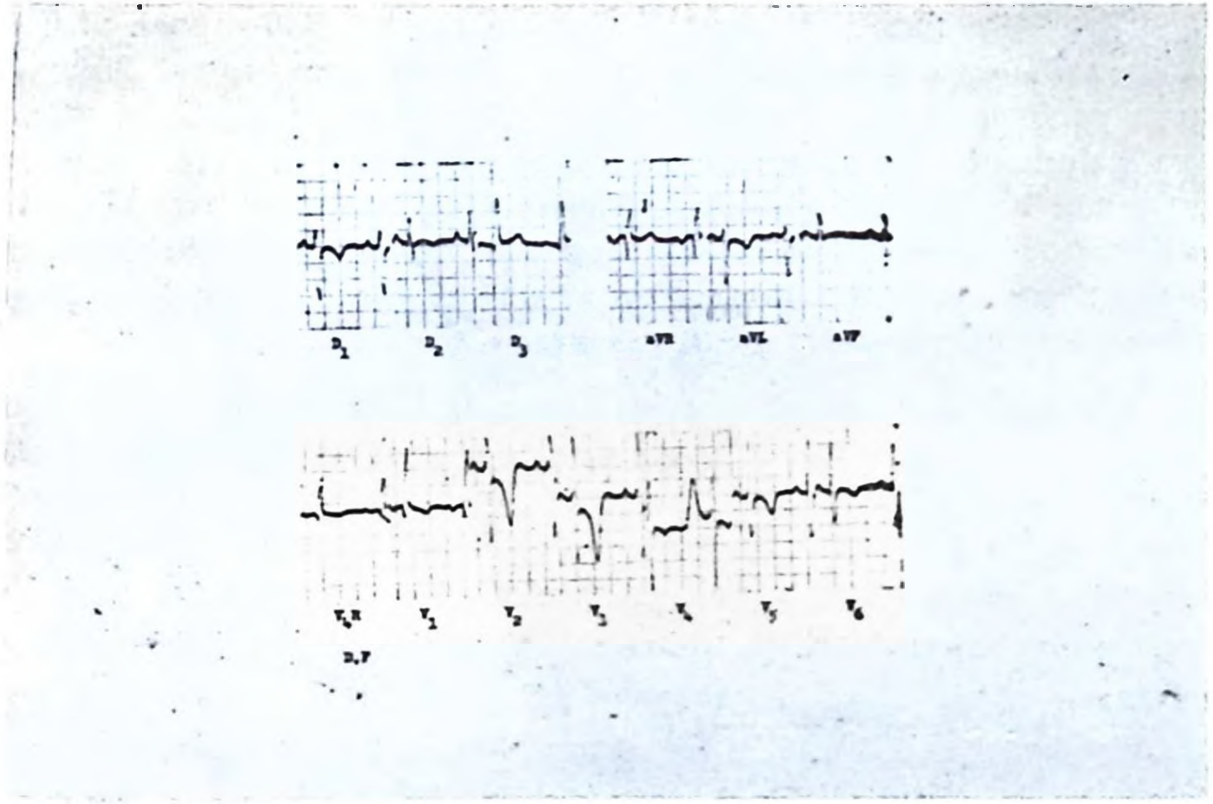

Fig. 9. Atypical electrocardiographic changes in tetralogy. Notice negative T waves in V_5 and V_6.

TABLE 3

AGE AND SEX DISTRIBUTION, AND SOME PROCEDURES EMPLOYED IN 100 SELECTED CASES OF TETRALOGY

Age Group	Number of Cases	Male	Female	Cardiac Cath.	Angio.	Only EKG and x-ray	Operation
Under 2	21	15	6	1	—	20	1
2 — 5	29	14	15	5	3	21	15
6 — 10	29	14	15	5	1	23	21
11 — 15	13	5	8	5	1	7	10
over 15	8	3	5	2	1	5	3
Total	100	51	49	18	6	76	50

The first column on the left in table 3 shows the age groups. In the group of children under two years of age we find a total of 21 patients, 15 of them male and six female. This might give the impression that tetralogy of Fallot is found twice as frequently in males as in females but if we look at the group as a whole we find that there are 51 males and 49 females, indicating that there are no significant sex differences in tetralogy.

The total number of cases of tetralogy of Fallot seen at Hacettepe Children's Hospital up to the present time is over 200. We have performed angiocardiography in 39 cases, only six of which are included in the group of 100 cases under review here. Since we have had two deaths due to angiocardiography we are now considerably more conservative about using this method with patients suffering from cyanotic or congestive heart disease. Unless it is absolutely necessary we prefer not to employ it.

Since cardiac catheterization can reveal only the probable presence of a patent foramen ovale or nonsaturation of oxygen in the systemic circulation and similar systolic pressure in the systemic artery and right ventricle and, since little supporting information to prove a diagnosis is provided by cardiac catheterization, we only use this technique in cases where the diagnosis is really challenging. Eighteen of our first 100 cases of tetralogy were catheterized but the number was reduced in the second 100 as our experience in making diagnoses by physical examination, EKG, roentgen ray and phonocardiography increased.

Surgical Intervention

Operations have been performed in the various age groups as follows : in the group under two years of age, only one operation; in the 2-5 year group, 15 operations; 6-10 year group, 21 operations; 11-15 year group, 10 operations; and over the age of 15 only three operations.

In all except three of these cases the Blalock operation was performed. The three exceptions were all under three years of age and all had the Potts operation. Our surgeons do a left thoracotomy regardless of the position of the aorta. If the pulmonary artery is wide enough and the subclavian artery can be anastomosed, they perform the Blalock operation. In children under two or three years of age, however, this is very difficult so the Potts operation is performed instead.

In all cases where the patient is under four or five years of age the small size of the main pulmonary and left subclavian arteries renders surgical intervention difficult so that operations are not performed on these children unless absolutely necessary. The indications and criteria in such cases are as follows : operations are performed when 1. a child past the age of two cannot walk, 2. a patient has an excessive number of cyanotic spells, 3. a child of three or more displays severe retardation of growth, or 4. the hematocrit is 70 per cent or more. In all other cases we prefer to wait until the child reaches the age of five.

Very few tetralogy of Fallot patients come to the hospital after the age of 15. We have had only eight such cases and performed operations in three of them. Most such cases are mild ones or involve patients who have already developed good collateral circulation, whose oxygenation is good, and who generally have less resistance in the pulmonary system. Operations were not considered necessary in five of these eight cases because the children rarely squatted and were able to walk about two miles. Squatting cyanosis was minimal at rest and the hemoglobin and red blood cell levels were not too high - perhaps slightly higher than the upper normal limit.

Table 4 shows the postoperative status of 50 cases. All of them had cyanosis; two of them died, one during the operation and the other right afterwards. Three patients expired several months after being operated on; one owing to diarrhea, one because of bronchopneumonia, and one for reasons unknown. Of the remaining 45 ca-

TABLE 4

POSTOPERATIVE STATUS OF 50 CASES OF TETRALOGY OF FALLOT*

Cyanosis 50 cases 2 deaths	32 none 5 less 3 same 5 ?	Clubbing 49 cases 1 death	6 none 13 less 1 same 25 ?
Murmur 50 cases 2 deaths	47 sys. - cont 1 only sys.	Dyspnea 46 cases 1 death	29 none 10 less — same 6 ?
Squatting 43 cases 1 death	30 none 6 less 1 same 5 ?	Palpitation 24 cases 1 death	17 none 5 less 1 same — ?
Striking Behavior Improvement in all Cases Except One.			

ses, cyanosis has disappeared in 32 cases, diminished in 5 and remained the same in three. In five cases the present degree of cyanosis, as compared with the pre-operative condition is not known. Several of these patients were unavailable for examination and the letters received about their present condition omit mention of cyanosis.

All the patients had systolic murmurs before surgery and these persisted after surgery. In addition, after surgery, continuous murmurs were found in all but one case. These continuous murmurs were variously located in the second left intercostal space, just under the left clavicle, and in the third intercostal space to the left of the sternum.

Squatting, which was reported by 43 patients before operation, disappeared in 30, diminished in six and remained the same in one. One patient died and there is no postoperative report on squatting in the other five.

Clubbing, which was present in 49 cases before surgery, was eliminated in six, diminished in 13 and remained the same in one. Four of these patients died, one during surgery and three later, and there is no report on the present degree of clubbing in 25 patients, owing in part to the difficulty of evaluating this condition and comparing it with the pre-operative situation.

* Three patients died several months after operation.

Dyspnea, present in 46 cases before operation, disappeared in 29 cases and was lessened in 10. One child died and there is no information on the other six.

Twenty-four patients complained of palpitations before surgery. Of these, 17 reported no palpitations after surgery, 5 had fewer and one remained the same. One patient died.

There was striking behavior improvement after surgery in all but one case. This was the same child who failed to show improvement with regard to squatting, clubbing and palpitations.

These children, before operation, were typically tense, nervous and touchy; refused food and gifts; rarely smiled; were suspicious of and hostile to the people around them; were manifestly miserable and cried with little or no provocation. Immediately after surgery, however, the picture changed dramatically. The children seemed to acquire new personalities, becoming aimiable and friendly with nurses, doctors, families and friends; smiling, accepting gifts, crying little; and generally evidencing confidence in their surroundings and the people caring for them... Are all these changes the result of better oxygenation of the brain following the shunt operation or should they be attributed to the greater freedom of physical activity permitted following surgery? Before their operations these children were not able to walk or play or participate in other children's ac-

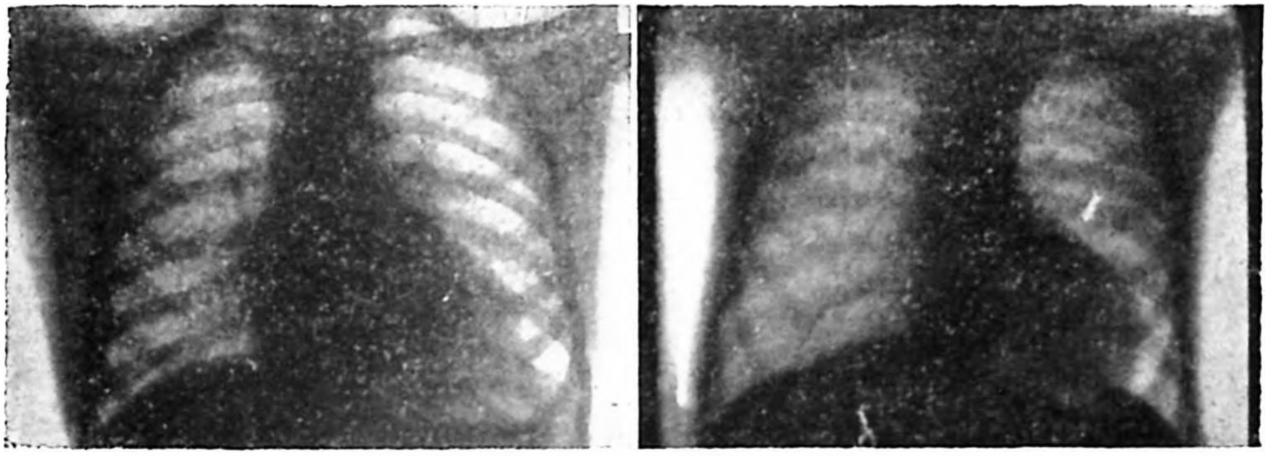

Fig. 10. Roentgenograms taken before (right) and after (left) the Blalock shunt operation in a patient with tetralogy.

tivities which might have tended to induce feelings of inferiority, distrust and depression. Perhaps this second explanation plays some part in the behavioral improvement but it is my feeling that better oxygenation of the brain plays the major role since marked personality and behavioral changes are manifest even before the children are allowed out of bed after surgery and when they are still in pain from the thoracotomy. In fact, lessened nervousness and greater aimiability are sometimes noted in the recovery room.

Postoperative Roentgen Ray Examination

Figure 10 presents roentgen ray films taken before and after surgery. On the right is a film taken before surgery and on the left is a film of the same individual taken six months after the shunt operation. The film on the right shows the typical tetralogy of Fallot picture : small heart, tip or apex elevated *(coeur en sabot)* and decreased vascularity in the lungs. Six months later the heart as a whole is larger and there is some increase in the vascularity of the lungs. The contour shows probable left ventricular hypertrophy.

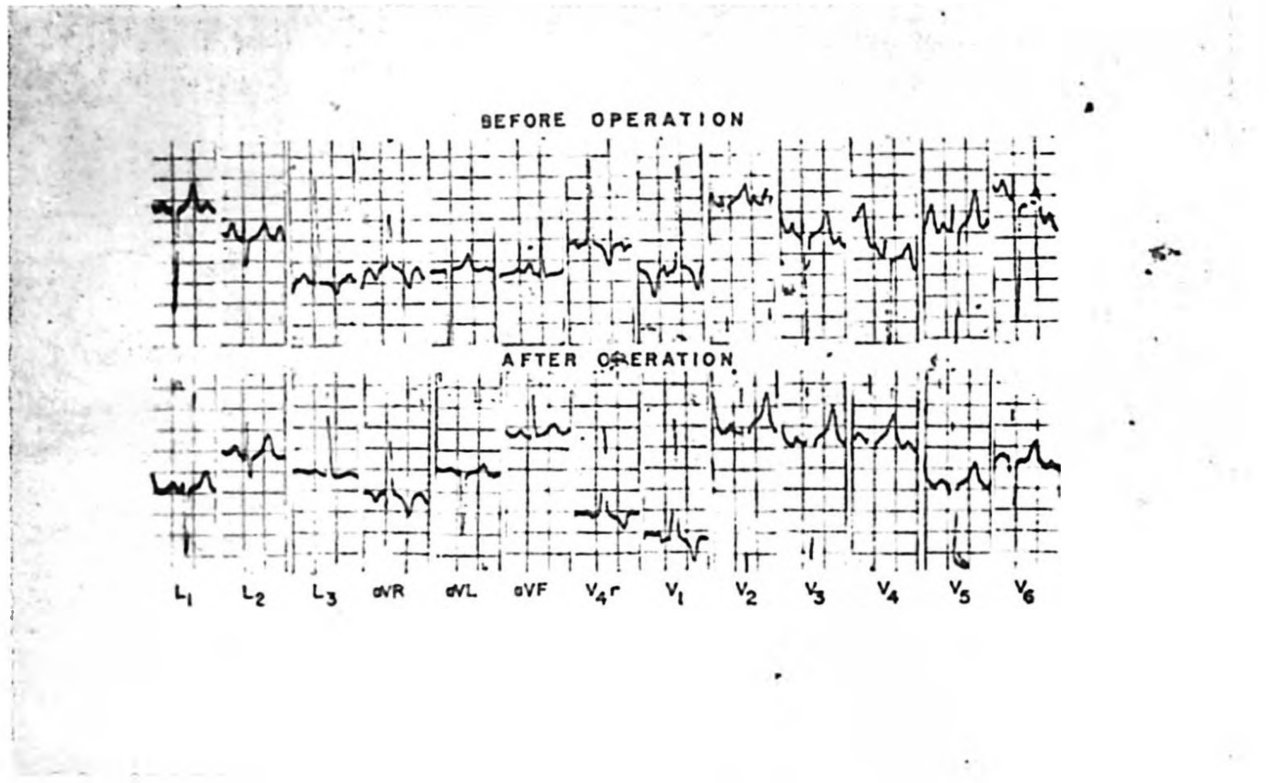

Fig. 11. Comparative electrocardiograms taken before and after the Blalock shunt operation.

Comparative Electrocardiograms

Figure 11 shows comparative EKGs before and after surgery. The EKGs done before the Blalock operation are in the upper row and those done six months after the operation are in the lower row. The standardization is the same in both.

An EKG done six to eight months after the Blalock operation shows striking changes. For example, as you see here, there were very small R waves in leads I and AVL before surgery whereas after surgery there is a definite increase in height in the R waves in those leads. The increase in the height of the R waves can best be observed in V5 and V6. Also notice the height of the R wave in V2.

Another common postoperative finding is the increased depth of the Q waves in AVR. A patient without Q waves in AVR before surgery, develops Q waves after surgery. A patient with Q waves before surgery shows deeper ones after surgery. The S waves too show changes, mainly in leads I, AVL, V5 and V6, becoming smaller after surgery. In some cases there are typical changes in P waves too, especially in leads I and II we find that the P waves are broader and that P waves which were not visible in AVL are now obvious. In some of our patients we have observed ST depression in the left precordium.

All these findings indicate that after surgery the left ventricle is somewhat overloaded and there is some evidence of the enlargement of the left atrium as well. If the EKG is done too soon after surgery it is not possible to see significant changes. In at least the first two weeks after surgery there is very little change and the significant changes are usually not obvious for several months.

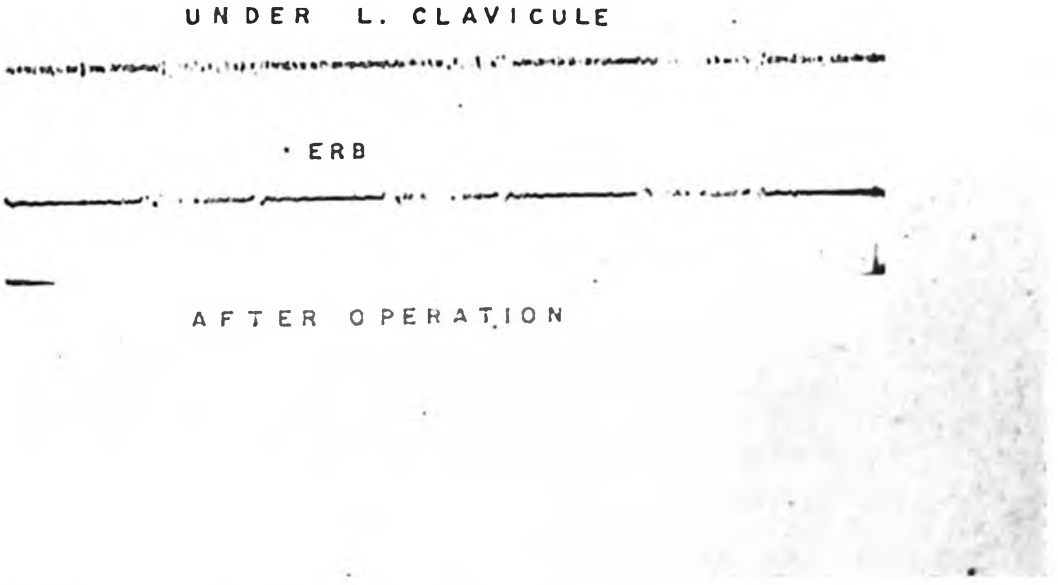

Fig. 12. Comparative phonocardiograms taken before and after the Blalock shunt operation. Note the obvious continuous murmur in the postoperative film.

In tetralogy cases with right ventricular hypertrophy before surgery we have never observed pure left ventricular hypertrophy without right ventricular hypertrophy after surgery. This is perhaps because we were doing mostly Blalock, not Potts, operations therefore there was little left ventricular overloading. Had we done

more Potts operations it is possible that we would have seen more left ventricular hypertrophy. On the basis of this EKG evidence it is perhaps possible to say that with long-term followup the Blalock operation may prove to be more satisfactory than the Potts since over a period of years patients having had the Potts operation may prove more prone to development of left ventricular heart failure.

Postoperative Phonocardiographic Changes

In figure 12 we see the postoperative phonocardiographic changes. The phonocardiogram on the top line was taken from under the left clavicle. The lower one was taken from the Erb. It is clearly seen here that there is an ejection type of systolic murmur at Erb and an obvious continuous murmur under the left clavicle.

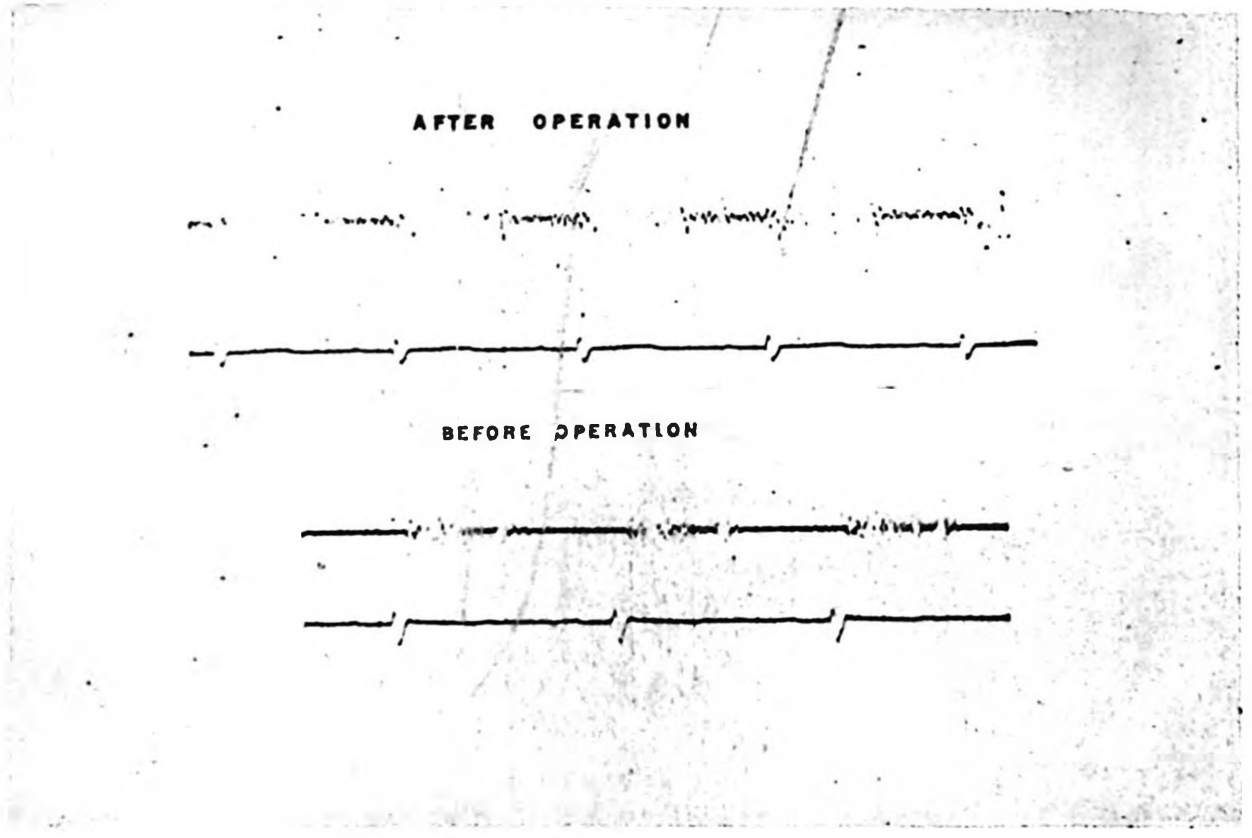

Fig. 13. Phonocardiograms taken six month before and six months after the Blalock shunt operation. The continuous murmur indicates that the shunt is functioning well.

Figure 13. These are phonocardiograms from another patient, the upper one taken six months after operation and the lower one six months before the operation. Both phonocardiograms were taken from the pulmonic area (second left intercostal space) with the same magnification, amplitude and microphone. It is obvious that there is a continuous murmur after surgery. We consider that the presence of this murmur indicates that the shunt is functioning well.

In table 5 we see the results of 50 operations performed on tetralogy of Fallot patients. In the first column the children are divided into five different age groups. The second, third and fourth columns show the degree of of success of the surgery. We considered the result excellent if cyanosis, squatting and adverse behavioral patterns had either completely disappeared or had, all three, shown significant improvement. The results were classed as showing moderate improvement if any of the three factors showed improvement or if all three were slightly improved.

TABLE 5

OPERATIVE RESULTS IN 50 TETRALOGY OF FALLOT CASES

Age Group	Excellent	Moderately Improved	No Change	Died	Total
under 2	—	1	—	—	1
2 — 5	7	4	1	3	15
6 — 10	17	3	1	—	21
11 — 15	6	2	1	1	10
over 15	2	1	—	—	3
Total	32	11	3	4	50

It can be seen from the table that only one patient under two years of age had been operated on and that this child was classed as showing moderate improvement. In the 2 - 5 year group excellent results were recorded in 7 cases, moderate improvement in 4, three patients died (two of them several months later) and one did not show any change at all. In the 6 - 10 year group we had excellent results in 17 cases, moderate improvement in three, one with no change, and no deaths. From 11 - 15 years we recorded excellent results in 6 cases, moderate improvement in two, and no change in one. One patient in this group died in the operating room under anesthesia before thoracotomy. Of those over 15, two showed excellent results and one, moderate improvement.

Taking the group as a whole we had excellent results in 32 cases, moderate improvement in 11, no change in three, and four deaths - one due to surgery, one to being anesthetized, and two to secondary causes.

The largest number of patients in any age group on which we operated was found between the ages of six and ten since it was

felt that the most favorable results from the shunt operation could be obtained in this age group. Twenty-one out of a total of 50 patients were in this group. A fairly large number of children (15) in the 2 - 5 year group were operated on but this was chiefly because they were severely handicapped by growth retardation, inability to walk, or frequent cyanotic spells. I should like to mention here in passing that we had one patient with severe symptoms of chorea who had been treated in another hospital for six weeks with phenobarbital, thorazine, and cortisone. The child showed no improvement at all with this treatment and displayed very exaggerated and agitated movements of the extremities. It was very difficult to keep her in bed or even to feed her. Immediately after the Blalock operation was performed it was noted that these exaggerated movements were significantly decreased and they disappeared entirely in a few days.

Summary

Common findings and symptoms in 100 selected cases of tetralogy of Fallot are presented together with the results of electrocardiographic, roentgenographic and angiocardiographic studies. Operative results in 50 cases are discussed and it is suggested that the Blalock operation, in preference to the Potts, may become the operation of choice in such cases in future.

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