BRONCHIECTASIS

A LONG-TERM FOLLOW-UP OF MEDICAL AND SURGICAL CASES FROM CHILDHOOD

BY

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Over the past 30 years the natural history of bronchiectasis must have been considerably modified by the varying treatments which have been introduced. Laennec's original description in 1829 was made from post-mortem material, the diagnosis in those early years only being made in severe clinical cases or at autopsy. With the introduction by Sicard and Forestier in 1922 of an opaque substance to outline the bronchial tree, more accurate diagnosis was possible, but whilst this new diagnostic technique was being improved, measures to combat the disease were introduced. Physiotherapy with postural drainage and breathing exercises was soon assisted by the introduction of chemotherapy and antibiotics. At about the same time surgical procedures, i.e. lobectomy and pneumonectomy, were adopted and later multiple segmental resections were practised in an attempt to remove, if possible, all the diseased parts. But it was soon realized that bronchiectasis was not just an anatomical dilatation of certain bronchi, but usually a general disease complex, and removal of all diseased parts did not necessarily relieve all the symptoms. As a result surgery was recommended less and less, particularly as chemotherapy was able to control the respiratory exacerbations.

The present survey of 225 patients observed over a period of eight to 21 years covers this period of changing conditions. In most cases the diagnosis was made before the introduction of antibiotics and in some cases before the use of sulphonamides. In spite of the varying treatments a definite pattern of disease seems to emerge in which the onset of damage to the lung is maximal in the first five years of life. Thereafter the symptoms may be troublesome until puberty, when a surprising improvement in health occurs, lasting usually through the 'teens into the twenties. It remains to be seen what happens in later life.

Earlier studies on these patients were reported in 1949 (Field) and the present study is a continuation of this work.

The Present Study

This report consists of 104 patients treated medically and 121 treated surgically. Although the author has been in Singapore for the past 11 years, contact with the patients was made on each home leave, and in 1956 137 patients were asked to attend for a clinical examination; 84 attended and 25, who were unable to attend, replied to a special questionnaire. There was no reply in 28 cases, usually because they had moved and the address was unknown. Out of these 109 patients, four were surgically treated and have been included in the surgical series and one who had died eight years previously was excluded, thus leaving 104 patients who constitute the medically treated patients of this study. All of these had at one time shown some dilatation of the bronchi by bronchogram, but in 38 instances this was mild and of doubtful permanency. All had symptoms of chronic chest disease.

Most of the surgical patients had been operated on by Professor R. S. Pilcher and his staff, and were being followed up regularly in his clinic. With his kind co-operation a special questionnaire was filled in by the patients when they attended the clinic and the examining doctor filled in the clinical record. A full record was obtained of 117 cases out of 150, and with four patients who had had operations from the medical clinic there were 121 surgically treated patients. In no way can this surgical series be compared with the medical series as selection for operation is essential, but the two records are given side by side. No attempt has been made to grade the severity of the bronchiectasis as this study is

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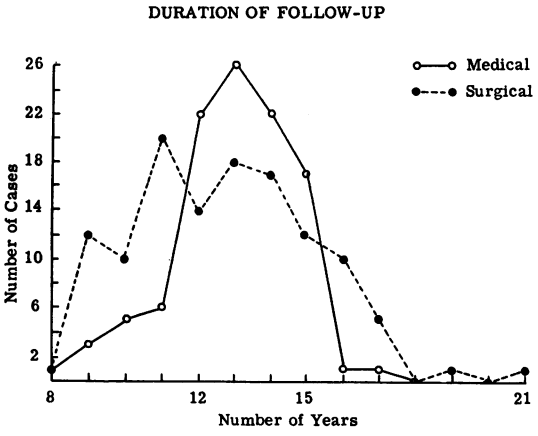

FIG. 1

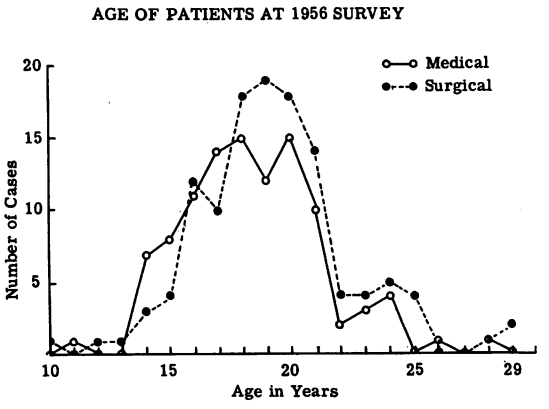

FIG. 2

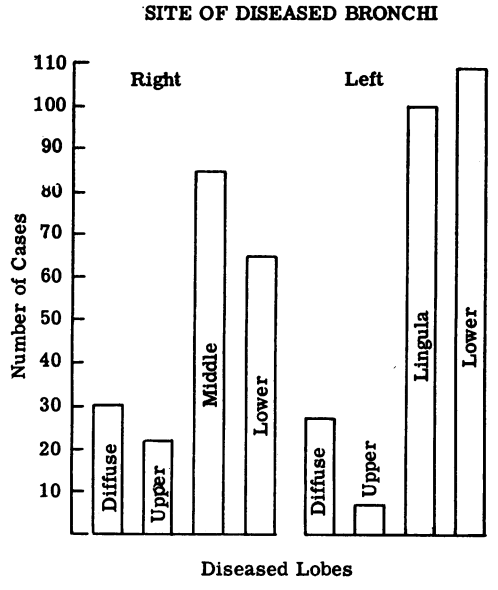

FIG. 3

TABLE 1

PERCENTAGE INCIDENCE OF TYPE OF BRONCHIECTASIS

Type	Per cent.
Tubular	35
Saccular	30
Varicose	27
Fusiform	11

more one of progress of the disease itself. There was no selection of cases at the onset; all cases who showed dilatation of the bronchi and who attended the children's Chest Clinics at two London hospitals over a period of several years were included. There were, however, different observers for the medical and surgical follow-ups.

Duration of Follow-up. It will be seen from Fig. 1 that the minimum number of years for which the patients have been followed is eight years and the maximum 21 years; the majority have been followed for 11 to 15 years.

Age. The majority of patients during the 1956 survey were between the ages of 14 and 24 years, the youngest being 10 years and the oldest 29 years (Fig. 2).

Sex. There are 102 males and 123 females; this supports the common finding of a slight preponderance of females.

Site of Disease (Fig. 3). As previously recorded (Field, 1949) the left lower lobe and lingula of the left upper lobe are most frequently affected, followed by the right middle lobe. In this series the right lower lobe is not quite so frequently affected.

Type of Bronchiectasis. Four types of bronchiectasis are described, but there are many variations of these and more than one type can be seen in one patient (Table 1).

Tubular dilatation is often the early stage of the disease in children, and this explains the high incidence. As the terminal part distends and becomes more bulbous, the bronchiectasis becomes

TABLE 2

AGE OF ONSET AND NUMBER OF CASES

Age of Onset	Number of Cases
First 12 months	50
1 year–5 years	119
5 years–10 years	51
10 years–15 years	3
No record	2

saccular in type. Varicose has been used here to describe the irregular dilatations along the length of the bronchus, and fusiform, the dilatation which is most marked in the middle of the bronchus (Field, 1949). The varicose and fusiform types usually have a good prognosis.

Children were only seen up to the age of 12 years in the chest clinics, but the figures in Table 2 show that the dangerous period is in the first five years of life, when 75% of the cases occurred.

Clinical Features. Although careful records were kept from the beginning of this study, it was only after several years that the importance of certain clinical features was recognized and recorded; the clinical records for these features, therefore, are inadequate for when the patients were first seen. However, where it is helpful, the records available will be shown. For the more important symptoms such as cough, sputum, nasal discharge, clubbing and moist sounds in the lungs, the early records are reasonably adequate, but where no record was available this is recorded.

Treatment. The medically treated patients received no special treatment; the majority had given up postural drainage and they were on no regular treatment. Antibiotics were usually given by their own doctors for exacerbation of chest symptoms, but as far as I know none of them received continuous chemotherapy.

TABLE 3

SITE OF PULMONARY RESECTION*

Site of Resection	Number of Patients	
	Right Lobe	Left Lobe
Upper lobes		
Complete	4	12
Segmental-antero-lateral	16	9
Segmental-apical	2	0
Segmental-postero-lateral	0	0
Middle and lingula lobes	67	72
Lower lobes		
Complete	20	56
Partial-anterior basic	16	7
Partial-middle basic	4	1
Partial-posterior basic	1	4
Partial-dorsal	10	8

* Multiple resections account for high total figures.

Pulmonary Resection. As might be expected, the lingula of the left upper lobe, the right middle lobe and the left lower lobe were the most frequently removed lobes of the lung; partial resection of a lobe was also performed not infrequently (Table 3).

Duration since Last Operation (Fig. 4). The majority of patients had their operation between three and 12 years ago, although three cases had a further operation only one year before clinical assessment.

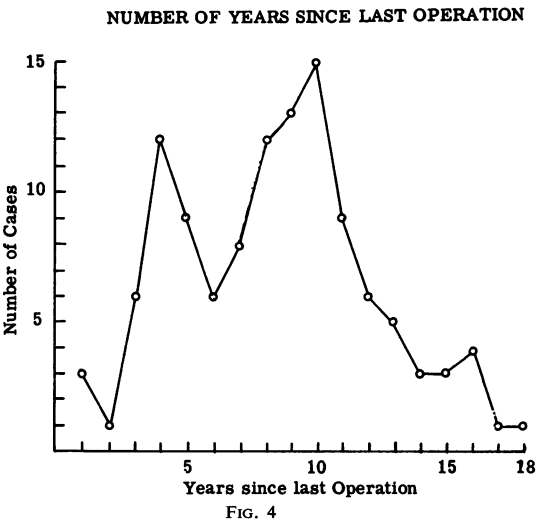

FIG. 4

Results of the Follow-up

Clinical Impressions. In the past, before statistical evidence was considered so necessary, great importance was attached to a good clinical description of a disease or disease process. At present clinicians find it necessary to prove their point statistically before it is accepted. Studying the present figures after the follow-up clinics in 1956 I realized how inadequate such figures were to express the clinical change that takes place in these bronchiectatic children. Therefore, before attempting to give the results of this investigation, I shall briefly describe my own clinical impressions of the disease process.

The bronchiectatic process most commonly commences in infancy as a result of chest infections, particularly pneumonia, measles or whooping cough; thereafter the child suffers from recurrent

bronchitis, often with an asthmatic wheeze, running nose and usually a persistently infected lung, as shown by a cough and moist sounds in the lung. At this age the disease process often seems to be generalized and is difficult to define. Recurrent pulmonary collapse is a common feature.

From 6 to 12 years of age the child's condition improves a little, although the cough usually persists often with a nasal discharge and sputum may be produced. Attacks of recurrent bronchitis or asthmatic bronchitis become less frequent and recurrent pulmonary collapse is less commonly seen.

As puberty approaches, a remarkable change seems to take place. No longer is the child anxious or worried about his disorder; many lose their cough and symptoms, and if they persist they cause little discomfort. Even their parents stop worrying about them. Their whole outlook on life has changed and their disease is quite a secondary matter. Clubbing usually disappears and moist sounds in the lungs are far less frequent. This is the picture of the mild and moderate case. The more severe cases are a little different. They usually persist with a cough which, however, becomes less troublesome; sputum may persist, but does not increase in quantity as might be expected, although some do complain of streaks of blood in the sputum.

On the whole the activity of the bronchiectatic adolescent is not impaired; most of them lead a reasonably full active life, but some complain that running makes them cough and they avoid it. The majority take jobs of one sort or another; most of these were either sedentary in nature or only required moderate exercise; but some preferred an open-air job. Many are now married, and none complained that their disease was a bar to marriage, although in a surgical case there has been one marital complication since marriage due to the danger of having further children. Reproduction appears to be normal, but growth as measured by height and weight does seem to be slightly impaired (see Figs. 6 and 7).

One small group in which diffuse bronchiectasis is associated with moderately severe asthmatic symptoms improve very little. Clubbing usually persists and the lungs are rarely clear of moist sounds; growth is stunted and there is often slight cyanosis showing the extent of lung damage, and they are unable to do a full-time job because of fatigue. The three cases in this category all died between the ages of 17 and 21 years from pulmonary insufficiency with signs of cor pulmonale. There have been eight deaths in the series since this study was first reported in 1947 (Field, 1949) and four more since 1956. These are reported in detail later.

Pulmonary resection undoubtedly improves certain cases if carefully selected for operation and relieves some of the most severe cases of many of their symptoms, but it does not seem to affect to any great degree the natural history of the disease. Bronchiectasis is so often something more than just localized damage to bronchi: it is a generalized respiratory disorder with a susceptibility to recurrent chest infections, sinusitis and their accompaniments, including allergy, and it is this that may persist in spite of surgical removal of the diseased parts. The patients treated surgically improved at puberty and during their 'teens, as did the patients treated medically, but they continue to have recurrent chest infections and there seems to be little to choose between the two groups.

Recurrent or persistent nasal infection and sinusitis is probably as troublesome to a bronchiectatic as his chest infection and it is often difficult to clear. This may account for some of the persistent symptoms.

Practically all the cases with recurrent pulmonary collapse showed, at a later date, typical asthmatic symptoms suggesting that the cause of recurrent pulmonary collapse is most commonly allergic in nature. Eight out of nine of these cases appeared to be quite well by puberty.

This story so far is only half told, for as the adolescent merges into adult life with marriage, children and responsibility some of the symptoms return and become troublesome, constituting what is commonly known as recurrent or chronic bronchitis in adults. As the follow-up continues it is my impression that the symptoms become more troublesome.

The exact part that *chemotherapy* and *antibiotics* play in the modification of the natural history of this disease is very difficult to assess. At the beginning of this study even the sulphonamides were not in general use and most of the cases were selected before chemotherapy could have had much influence. However, there is no doubt that antibiotics play an important part in the recurrent chest infections and reduce the dangers of complications in surgery so that their use must have affected to some extent the prognosis; it is doubtful if it has affected to any great degree the prevention of recurrent chest infections. As far as I know continuous chemotherapy has not been used on any of these children. It seems, therefore, that the natural history of bronchiectasis continues as before with modifications due to the use of antibiotics during respiratory exacerbation, which probably slows down quite considerably the deterioration that may occur in the more severe cases. The use of anti-

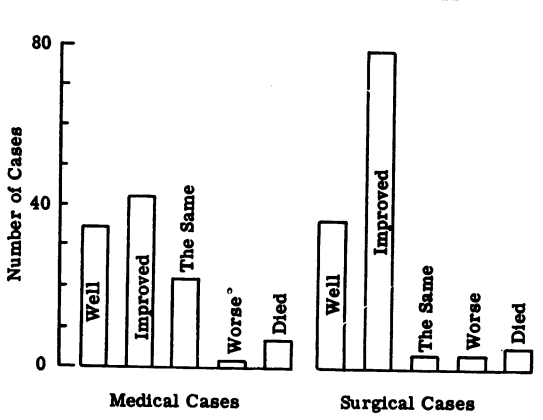

FIG. 5

biotics is probably not responsible for the great improvement seen at puberty in this series of cases; it seems that this has been and is the natural process of the disease. I am unable to give a satisfactory explanation, but there are three possibilities:

(*a*) An endocrine influence.

(*b*) Anatomical growth of bronchi and muscles, resulting in better expulsive power and drainage.

(*c*) Psychological: the adolescent usually has so many absorbing interests that health becomes of secondary importance; they also tend to look at life through rose-tinted spectacles.

I do not think (*c*) can account for all this change which is probably a combination of many factors.

Bronchiectasis in association with tuberculous hilar glands is often slow in developing and causes fewer symptoms. It is usually in the upper lobes when it is doubtful if pulmonary resection is justifiable as symptoms are few or absent. When it occurs in the middle or lower lobes and produces symptoms, pulmonary resection is advisable. It seems that bronchiectasis following tuberculosis is less troublesome.

Analysis of the Results. In an attempt to prove the clinical impressions just recorded, the following data were compiled from the records kept in 1956 of 104 patients treated medically and 121 patients treated surgically, who were followed up for eight to 21 years.

General Assessment (Fig. 5 and Table 4). When all the data had been collected a general assessment of progress was made by the doctor.

Only four children have deteriorated and four have died, although the condition of 22 medically

TABLE 4

DATA FOR FIG. 5

Assessment of Progress	Number of Cases	
	Medical	Surgical
Well	35	36
Improved	42	79
The same	22	3
Worse	1	3
Died*	4(+3*)	0(+5*)

* See under Deaths.

treated patients, as opposed to only three surgically treated patients, remained the same. The number of deaths gives a deceptive impression as those children known to have died before the 1956 survey were not included in the survey, but are fully reported later in this paper and included in Fig. 5.

Intercurrent Illnesses (Table 5). Bronchitis is the most frequent and troublesome complication which usually, however, responds well to antibiotics. About 50% of the patients have suffered from an intercurrent chest disorder.

TABLE 5

NUMBER OF PATIENTS SUFFERING FROM INTERCURRENT ILLNESS DURING THE PAST SIX YEARS

Illness	Patients Treated Medically	Patients Treated Surgically
Pneumonia	14	8
Bronchitis	28	47
Asthma	14	8
Miscellaneous	33	22
None	39	19*
No record	3	26*

* Surgical patients with no illness were often not recorded so that many cases under 'No record' could really be under 'None'.

Time Off Work or School (Table 6). Twelve medical patients with over 16 weeks' sick leave indicate that the more severe cases are incapacitated; three of these cases died shortly after these records had been collected. In about 50% of the

TABLE 6

NUMBER OF WEEKS SICK LEAVE DURING THE PAST SIX YEARS (EXCLUDING OPERATIONS)

Sick Leave (weeks)	Patients Treated Medically	Patients Treated Surgically
None	27	21
1–2	27	38
3–4	17	23
5–8	13	11
9–16	4	14
>16	12	6
No record	4	8

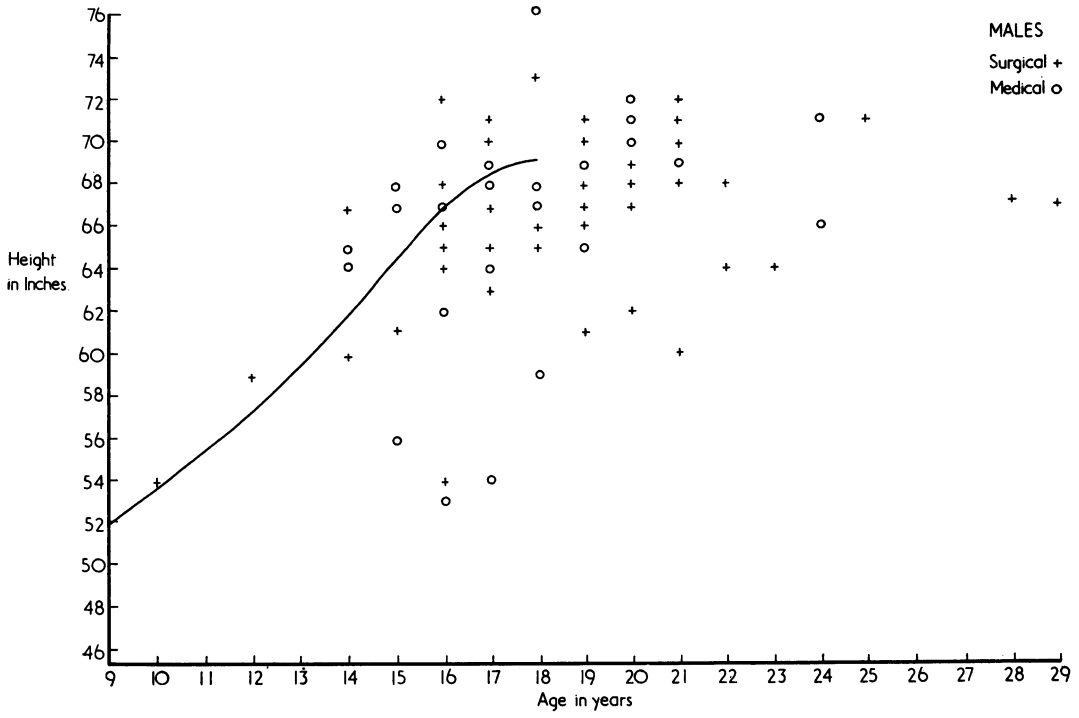

FIG. 6a.—Heights in males related to 50th percentile.

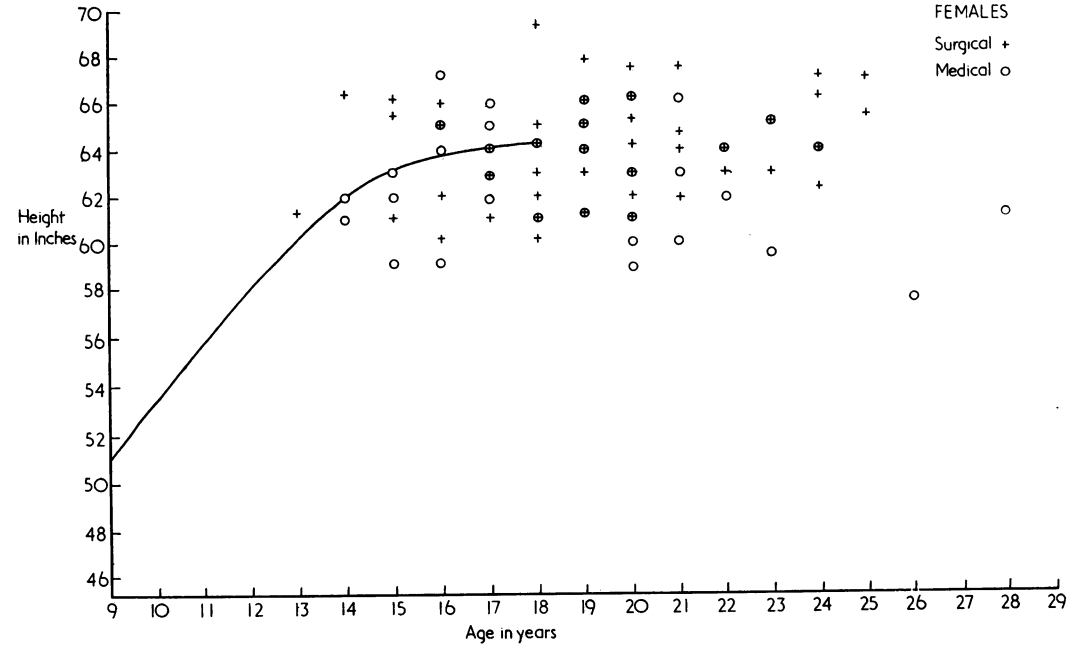

FIG. 6b.—Heights in females related to 50th percentile.

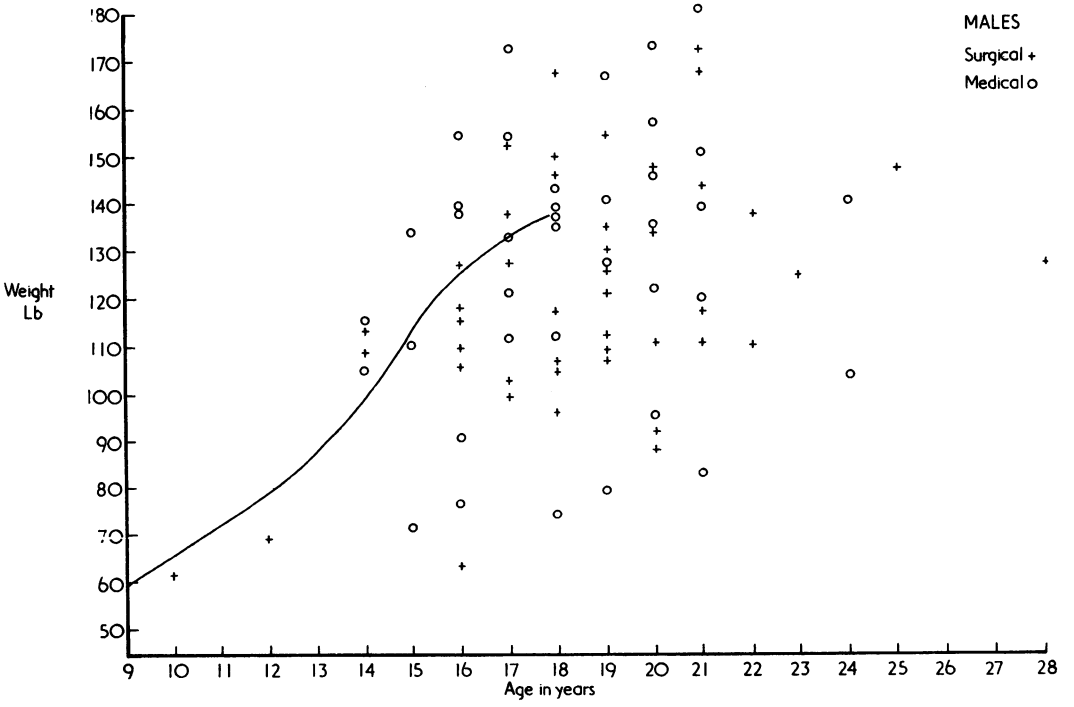

FIG. 7a.—Weight in males related to 50th percentile.

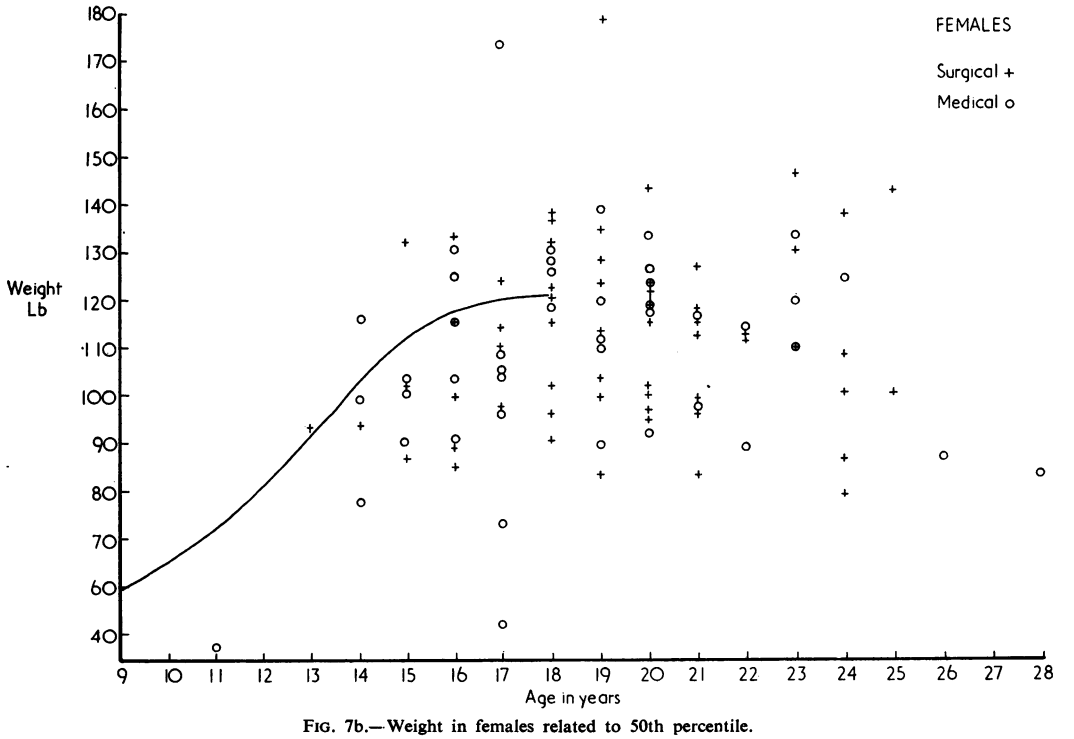

FIG. 7b.—Weight in females related to 50th percentile.

TABLE 7

PATIENT'S ASSESSMENT OF HIS HEALTH DURING THE PAST SIX YEARS

Assessment	Patients Treated Medically	Patients Treated Surgically
Improved	84	93
The same	12	19
Worse	3	3
No record	5	6

cases the cause for sick leave was connected with the chest.

Patient's Assessment of His Health (Table 7). Each patient was asked whether he felt his condition had improved over the past six years or was the same, or whether it was worse.

This assessment is similar to the evaluation of the doctors shown in Fig. 5; the vast majority say they feel better.

Marriage. When these records were compiled in 1956, nine medical and 14 surgical patients were married with an aggregate number of 18 children between them. The health since marriage of 11 had improved and of 10 was the same; nobody was worse, but there was no record for two. One patient now living in Canada informs me that the doctor has advised her against having further children and recommends sterilization. This has caused difficulties in the family. The patient, now aged 29 years, always rather fragile, was one of the early surgical cases in which the right middle lobe, left lower and lingula lobes were removed, and the left upper lobe failed to expand and finally had to be removed. She has therefore reached the age of 29 years (at the time of writing), has been married for eight years, and has had one child, with only the upper and lower lobes of the right lung.

It is too early at this stage to give a report on the effect of marriage in these patients. Many more are now married and further records may be revealing.

Clinical Assessment. Although careful records were kept at the beginning of this study, the importance of certain clinical features was not realized till later and the records are incomplete. Usually, if no record was made, it meant the symptom was not present, but this cannot be assumed. In the important features the records are reasonably complete and will be recorded.

Growth and Nutrition. Figs. 6 and 7 show the individual records for height and weight in relation to age and in relation to the average 50th percentile. It will be noted that more patients in all charts are below the 50th percentile than above it and some patients, particularly those treated medically, are quite severely undersized. It had been noted that the severe cases, particularly those with asthma, were stunted, but the general tendency for the bronchiectatic to be undersized was a surprise finding.

Cough. Fig. 8 and Table 8 show clearly that the severity of the cough has improved over the years and 44 medically treated patients and 31 surgically treated patients are now free from cough. When it is present it is far less persistent than in childhood.

Sputum. Unfortunately early records are incomplete, but data in Fig. 9 and Table 9 show a tendency to lessening of the amount of sputum. Haemoptysis was rare in infancy, but streaking of sputum with blood was noticed in a few patients during adolescence. One patient died of a severe haemoptysis.

Nasal Discharge. Two records were kept of nasal discharge: the patient's complaint and the doctor's observation. The simple clinical test of blocking each nostril separately with the finger and asking the patient to sniff was applied.

Figs. 10 and 11 and Tables 10 and 11 show that there was a definite improvement in the nasal discharge during adolescence both as a symptom and as a clinical sign. Many of these children had sinus operations in childhood. It seems that the sinus infection is as troublesome as the chest infection in many of these patients and probably accounts for some of the sputum.

Wheezing. In children this is often associated with respiratory infection and seems to be less marked as the child grows older. Early records are incomplete, but the 1956 records (Table 12) show that a number still suffer from this complaint occasionally.

Breathlessness. This is a vague and ill-defined symptom and early records are incomplete. In the 1956 survey there were many, particularly in the surgical group, who said they became breathless on exercise (Table 13). This is of no significance without further tests, but is something which might be investigated.

Clinical Signs. For those patients who were unable to attend for a clinical examination but replied to a questionnaire, no clinical records were obtained. However, one patient had been examined

SEVERITY OF COUGH
(Combined Medical & Surgical Cases)

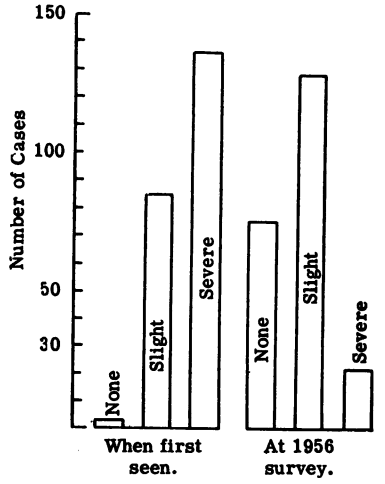

FIG. 8

TABLE 8

DATA FOR FIG. 8

Severity of Cough	Number of Cases		
	When First Seen	1956 Survey	
		Medical	Surgical
None	3	44	31
Slight	83	47	81
Severe	136	12	9
No record ..	3	1	0

AMOUNT OF SPUTUM
(Combined Medical & Surgical Cases)

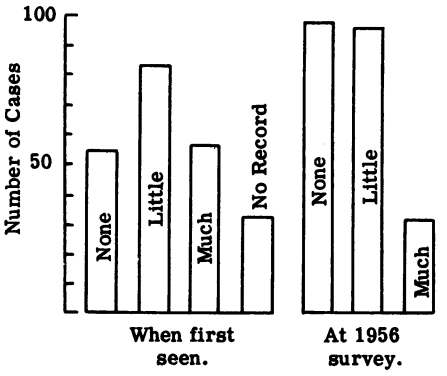

FIG. 9

TABLE 9

DATA FOR FIG. 9

Amount of Sputum	Number of Cases		
	When First Seen	1956 Survey	
		Medical	Surgical
None	55	48	50
Little	82	38	58
Much	56	18	13
No record ..	32	0	0

NASAL DISCHARGE
AS A COMPLAINT BY THE PATIENT

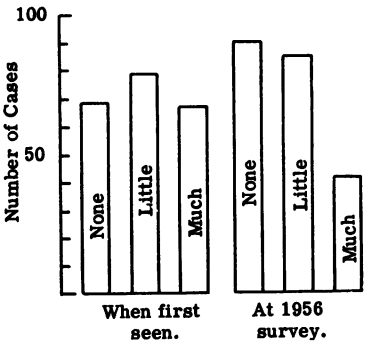

FIG. 10

TABLE 10

DATA FOR FIG. 10

Complaint of Nasal Discharge	Number of Cases		
	When First Seen	1956 Survey	
		Medical	Surgical
None	69	47	43
Little	79	33	52
Much	67	21	20
No record ..	10	3	6

two years previously and 14 patients three years previously, and these clinical records were accepted. In a few patients a clinical report was obtained from the private practitioner.

Early records are incomplete except for clubbing and moist sounds in the lungs.

Posture, Shape of Chest and Chest Expansion. Postural defects are not uncommon, particularly round shoulders, protuberant abdomen and lordosis, and these do not seem to improve during

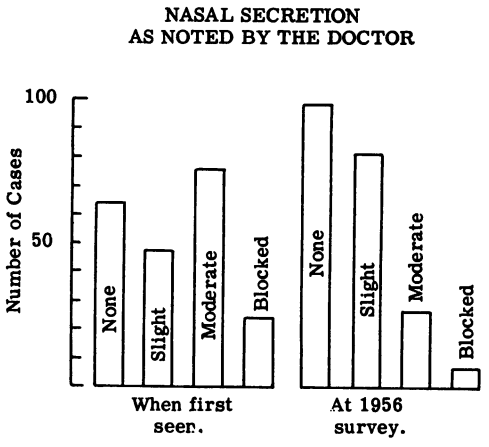

FIG. 11

TABLE 11

DATA FOR FIG. 11

Observed Nasal Secretion	Number of Cases		
	When First Seen	1956 Survey	
		Medical	Surgical
None	63	44	55
Slight	48	35	48
Moderate	76	13	14
Blocked	23	5	2
No record	15	6	5

TABLE 12

WHEEZING AS A SYMPTOM IN THE FOLLOW-UP RECORDS OF 1956

Symptoms of Wheezing	Patients Treated Medically	Patients Treated Surgically
None	60	65
Little	32	43
Much	10	10
No record	2	3

TABLE 13

BREATHLESSNESS AS A COMPLAINT IN THE FOLLOW-UP RECORDS OF 1956

Breathlessness	Number of Patients	
	Medical	Surgical
None	50	45
On exercise	48	71
At rest	3	2
No record	3	3

adolescence. Postural defects after pulmonary resection, particularly curvature of the spine and flattening of one side of the chest, may also persist. Sometimes flattening of the chest follows pulmonary collapse. Pigeon chest was most commonly seen in those children with associated asthmatic symptoms. Tables 14 and 15 illustrate the prevalence of chest deformities.

Chest Expansion (Table 16). The greater number with 'good expansion' in the surgically treated patients is probably the result of breathing exercises which are practised more diligently after surgery. Very few medically treated patients persisted with the breathing exercises.

Clubbing. In bronchiectasis, clubbing usually indicates a purulent infection of the chest. With the use of antibiotics the infection has come under control and the clubbing has disappeared, as it has also after surgical removal of the diseased part. Fig. 12 and Table 17 show the disappearance of clubbing in about one-quarter of the cases.

Moist Sounds in the Chest. Moist sounds in the lungs are usually indicative of some persistent infection or allergy. Fig. 13 and Table 18 show the tremendous improvement in both medically and surgically treated patients in adolescence; in 129 patients no moist sounds were heard in the follow-up survey.

At the onset of the disease râles were more in evidence than rhonchi, and in the follow-up they were of equal prevalence.

General Comments on Clinical Features. These records support the clinical impressions of a subjective and actual improvement in the average case of bronchiectasis during adolescence. The cough, sputum, clubbing and moist sounds in the chest all improve, although nasal secretion continues to be troublesome and posture may remain defective.

Bronchographic Changes. Repeat bronchographic studies have not usually been done on these children during adolescence; nevertheless, certain changes have been observed over the years in specific cases.

As already stated, the fusiform and varicose types of bronchiectasis usually have a good prognosis. The saccular type is variable, depending on the extent of the disease and the associated damage to the bronchi. The patient continues to have a cough with sputum and some of these have persistent clubbing. The tubular type of bronchiectasis shows definite changes over the years and in itself it is

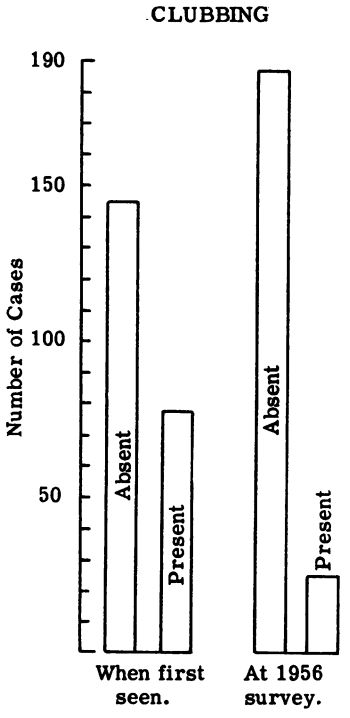

FIG. 12

probably not a stable form. Three changes have been noted.

1. The bronchi revert to normal calibre with the re-expansion of a pulmonary collapse (Fig. 14a and b).
2. The peripheral end of the bronchus becomes

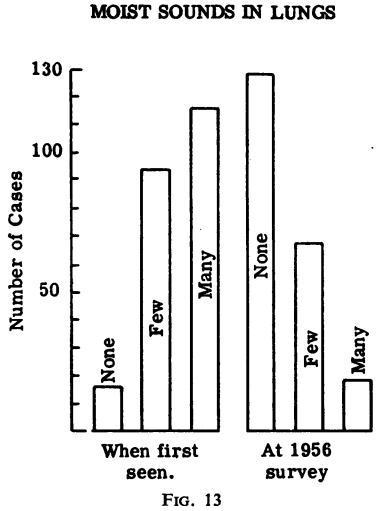

FIG. 13

TABLE 14

POSTURE IN PATIENTS SEEN IN 1956 SURVEY

Posture	Number of Patients	
	Medical	Surgical
Good	69	58
Slight defect	18	39
Bad	5	20
No record	12	4

TABLE 15

SHAPE OF CHEST IN PATIENTS SEEN IN 1956 SURVEY

Shape of Chest	Number of Patients	
	Medical	Surgical
Normal	67	54
Pigeon	10	13
Flattened	16	42
Spinal curve	6	16
No record	10	5

TABLE 16

CHEST EXPANSION IN PATIENTS SEEN IN 1956 SURVEY

Chest Expansion	Number of Patients	
	Medical	Surgical
Good	21	57
Average	55	48
Poor	11	10
No record	17	6

TABLE 17

DATA FOR FIG. 12

Clubbing	Number of Cases		
	When First Seen	1956 Survey	
		Medical	Surgical
Absent	145	86	101
Present	78	12	12
No record	2	6	8

TABLE 18

DATA FOR FIG. 13

Moist Sounds in Lungs	Number of Cases		
	When First Seen	1956 Survey	
		Medical	Surgical
None	16	55	74
Few	93	29	39
Many	116	15	4
No record	0	5	4

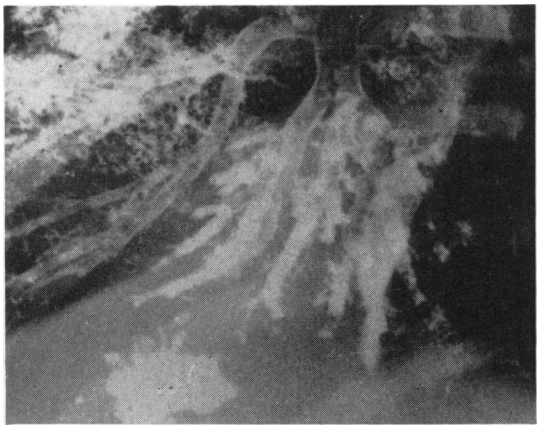

FIG. 14(*a*).—Left oblique bronchogram of a girl aged 7 years, showing the dilated lower lobe bronchi and the lingula lobe bronchi displaced downward and slightly dilated. The patient gave no history of cough only loss of weight and she easily became tired. Pulmonary collapse of the left lower lobe was revealed on a radiograph two months previously.

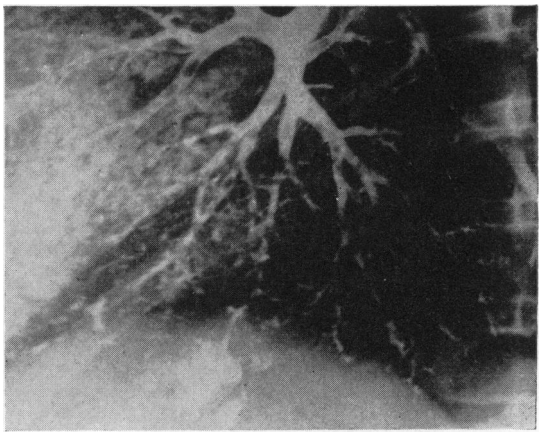

FIG. 14(*b*).—Left oblique bronchogram of the same patient repeated after one year and nine months. The dilated bronchi are now normal width.

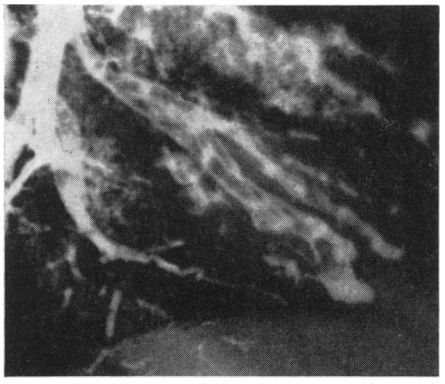

FIG. 15(*a*).—Right lateral bronchogram of a girl aged 4 years 9 months, showing the right middle lobe bronchi with tubular dilatation.

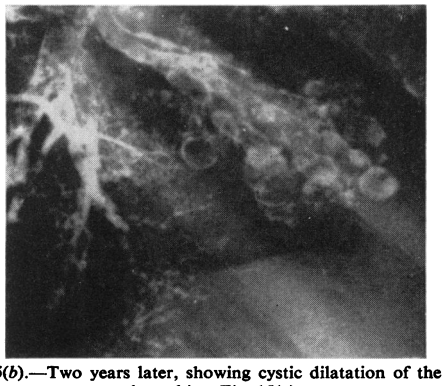

FIG. 15(*b*).—Two years later, showing cystic dilatation of the same bronchi as Fig. 15(*a*).

bulbous and often cystic (Fig. 15a and b). This is a sign of irreversible bronchiectasis.

3. The tubular dilatation takes on a varicose appearance (Fig. 15c).

In infants the tubular dilatation is often diffuse and difficult to define, but over the years it so commonly localizes itself to the dependent bronchi; in those bronchi in which gravity assists drainage the dilatation clears up. This accounts for the common occurrence of the distribution for bronchiectasis, i.e. the basal branches of the lower lobes, the right middle and lingula lobes and the anterolateral branch of the upper lobes when the middle and lingula are affected. (When bronchiectatic,

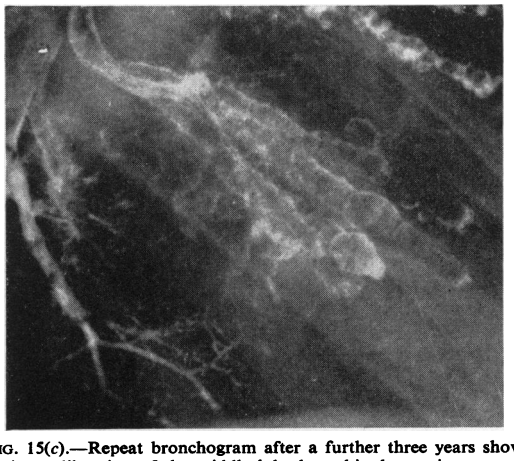

FIG. 15(*c*).—Repeat bronchogram after a further three years shows varicose dilatation of the middle lobe bronchi; the cystic areas are not filled but can be seen on the original film.

the middle and lingular lobes take up a smaller space, displacing the antero-lateral branch of the upper lobes downwards.) The upper lobes and dorsal part of the lower lobes commonly escape. This shows the great importance of adequate drainage in the prevention and treatment of early bronchiectasis. After lobectomy of the middle and lower lobes, some of the upper lobe bronchi become dependant as the lobe re-expands to occupy a large space, and if these bronchi are in any way diseased they will become bronchiectatic. If the bronchi are healthy the lung expands well and the bronchi often appear to grow in length ultimately occupying a similar space to the whole lung before removal. This phenomenon is seen most strikingly in the younger child.

Bronchiectasis in association with tuberculous lesions develops slowly even when associated with tuberculous hilar glands. After many years it may still not be present if secondary infection has been prevented.

Deaths. Since the original publication (Field, 1949) when 19 deaths were reported out of 202 cases, 12 more patients have died. Seven of these had no operation. Only four died near enough to the 1956 survey to include in the clinical assessment. Table 19 gives an analysis of these deaths. Cases 1 and 2 were young children having had pulmonary resection some time previously, but whose general condition and pulmonary state progressively deteriorated; both patients were then suspected of suffering from fibrocystic disease of the pancreas, but this was never proved. Case 3 was observed from the onset when she developed cystic bronchiectasis after a virulent staphylococcal pneumonia which had been treated in the early days with sulphathiazole (Fig. 16a and b). Her condition was never satisfactory after this, and, in the final stages, her private practitioner found massive collapse of the left lung and extensive infection of the right. Case 4 was a child with an obscure myopathy, always frail and fragile. She died in hospital with a severe haemop-

TABLE 19

ANALYSIS OF DEATHS

Case No.	Age at Death (years)	Sex	Extent of Bronchiectasis	Pulmonary Resection	Cause of Death	Autopsy
1	10	M	Bilateral, extensive	Right mid lobe and segmental right upper lobe Segmental right lower Segmental left lower	Suspected fibrocystic disease of pancreas not proved	No
2	10	F	Bilateral, extensive	Left lower lobe	Died in cardiac failure with cor pulmonale; also suspected fibrocystic disease of pancreas not proved	No
3	17	F	Bilateral, extensive after staphylococcal pneumonia	None	Left lung later collapsed and condition rapidly deteriorated; probably died of pneumonia	No
4	18	F	Bilateral, diffuse; also suffered from myopathy	None	Pneumonia; severe haemoptysis	No
5	19	F	Bilateral, diffuse with severe asthma	None	Died in cardiac failure with cor pulmonale; pulmonary oedema and bronchitis; lung abscess and generalized amyloidosis	Yes
6	20	M	Bilateral, diffuse with asthma	None	Died at home in cyanotic attack; radiograph showed cor pulmonale	No
7	21	M	Bilateral, diffuse with asthma	None	Doctor's report 'died at home with a story that sounded like cor pulmonale with ascites and oedema'	No
8	7	M	Bilateral, cystic	Left lower lobe Segmental right upper lobe	Died from oedema of lungs three days after second operation	Yes
9	11	F	Cystic right mid and lower lobes; scattered cysts in left lung	Right middle lobe	Died from toxaemia and renal failure 18 days after operation	No
10	6	F	Extensive	Left lower lobe	Died five years after operation with extensive bilateral cystic bronchiectasis	Yes
11	16	F	Cystic right upper lobe	None	Died in uraemia from chronic renal disease	No
12	23	M	Tubular right mid lobe and lingula	None	Died in motor cycle accident	No

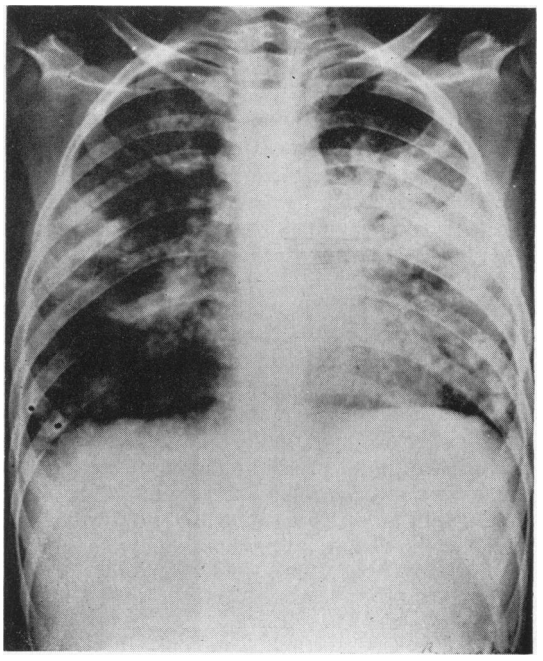

FIG. 16(*a*).—Antero-posterior radiograph of chest of a girl aged 10 years with staphylococcal pneumonia.

tysis after a recurrence of her chest infection. Cases 5, 6 and 7 suffered from extensive bilateral bronchiectasis with moderately severe asthma; the radiographs showed cor pulmonale and also marked emphysema. Cases 8 and 9 died in the early days of surgery. Case 10 was a patient with extensive bilateral cystic disease who died five years after removal of the left lower lobe. Cases 11 and 12 did not die as a result of their bronchiectasis.

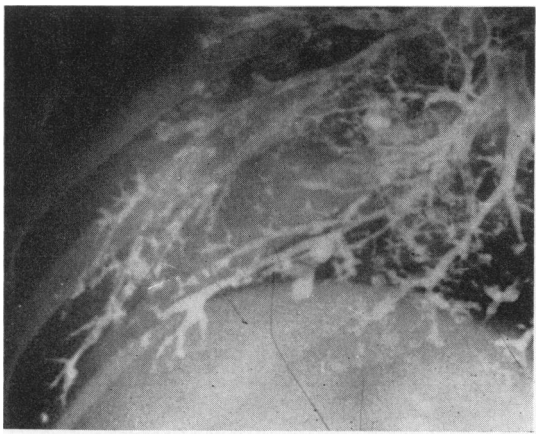

FIG. 16(*b*).—Left oblique bronchogram of the same patient three years later, showing part of the left lower and lingular bronchi with small cystic dilatations, which were present diffusely over both lung fields.

It is the first three cases that are perhaps puzzling: the children whose bronchiectasis never seems to respond satisfactorily to treatment. There are a few of these cases still living, and, as can be seen, surgery does not help. The extent of the lung damage, the virulence of the infection, or an underlying general disorder such as fibrocystic disease of the pancreas or agammaglobulinaemia should be suspected.

Discussion

In the past 10 years there have been several follow-up studies reported in the literature both for medically treated and surgically treated cases, and on the whole the results are favourable with the majority showing improvement and with some cases apparently clinically well (McKim, 1952; Strang, 1956; Dyggve and Gudbjerg, 1958; Franklin, 1958). The only study, however, that can be compared with the present series is that by Strang. He followed up 209 children all under 15 years at the time of onset of the disease; the duration of follow-up was shorter, two to 15 years. In 75% of his cases the onset of the disease was in the first five years of life; 51% showed clubbing and his cases may be, on the average, more severe than the series reported here. One hundred and sixty-three were treated surgically and 46 conservatively. There is no doubt that Strang was seeing the same trend of improvement and in fact the results in his surgically treated patients are very similar to those reported here. The prognosis in his medically treated patients is not quite so good, but this may be because, in the present series, 38 cases were mild cases, some of which were of doubtful permanency, and many of these are now symptomless and probably cured. Strang mentions the frequency of feverish chest illnesses which occurred in 130 of his 209 cases. It is these febrile attacks that can now be effectively controlled in most patients with antibiotics, thus changing to a certain extent the natural history of the disease.

Ogilvie, in 1941, records the results in 68 cases of bronchiectasis. Although these were followed for 'less than 6 years', nevertheless, they occurred in the era before the extensive use of antibiotics. Even in these cases 25 were well and symptom-free; 16 of these, however, were after operation. Fifteen had died, six after operation. Some of his cases, however, included adults as the average age was 17 years 3 months. Strang (1956) mentions a further follow-up for 11 to 18 years of 15 of Ogilvie's bronchiectatic children; five had died, two were

'cured', three had greatly improved and five remained the same but carried on a normal life.

McKim (1952) reviews 49 ambulant cases collected between 1930-41, which were followed up for nine to 20 years. The average age at the time of diagnosis was 31 years, so this series was, in the main, from a different age period. Nevertheless, he found that there was a general tendency to improvement, but that the longer the history of the disease the less good was the prognosis. Does this suggest that all cases of bronchiectasis improve for a time only or is it a question of the age factor?

Franklin (1958) divides his 20 patients into three categories: invalid, delicate, and absolutely well after the first assessment in 1944. In 1958 these patients had been followed for six to 22 years and their ages ranged from 15 to 34 years. The 11 patients who were absolutely well remained well; of the four delicate patients, two remained delicate and two became invalid, and of the five invalid patients, one had died, one had become well and the others had remained invalid or delicate. This second review suggests a slight overall deterioration, but on the whole there is not much change. Many of these patients are now in their twenties; only one is in the thirties.

In the series of cases reported here, there has been no classification of severity, largely because the review is essentially an overall review relating to the natural history of the disease process. Nevertheless, it is possible to study those patients who either died or whose condition has worsened. Strang (1956) states 'there was no evidence that the severity of the symptoms before operation had any bearing on the ultimate result'. With that I am inclined to agree as removal of a severe but localized cystic lobe can produce a symptomless patient. I have seen the same improvement occur in a severe case of bilateral saccular disease where operation was refused. The disease must have become 'dry' as, at the age of 16, the child had no cough or sputum and was stated to be well. Such a case, however, is a rarity; in general, those who are treated conservatively, may improve but continue to be severe. The mysterious cases are those who for no apparent reason deteriorate rapidly in spite of treatment by antibiotics. Case 3 (Table 19) was such a patient. Her fine diffuse cystic bronchiectasis following staphylococcal pneumonia did not look severe (Fig. 16a and b), yet her progress was never satisfactory; a superadded massive collapse of the left lung must have decided the ultimate outcome. It is probable that the disease in the lung was more extensive than the bronchograms suggested and that after the massive collapse she was left with inadequate pulmonary tissue for gaseous interchange. This is one of the most dangerous signs in cases of bronchiectasis usually manifesting itself with some degree of cyanosis. A further case illustrates this, a brother and sister were both affected with a somewhat diffuse type of bronchiectasis in childhood. The sister made good progress, but the brother never seemed well. Lobectomy was carried out and the most severely diseased parts were removed, but this produced little or no improvement; he has now reached the stage where he is unable to work due to fatigue and has cyanosis and breathlessness on exertion. The damage to the pulmonary alveoli in this patient is too extensive for satisfactory ventilation and in due course circulatory embarrassment will follow. These children have a bad prognosis, particularly as they reach adolescence and their physiological needs are greater. From Table 19 it can be seen that Cases 5, 6 and 7 are of this nature and probably Cases 1, 2, 3 and 4. Operative procedure on such cases is risky if the margin of healthy pulmonary tissue is small. Where progress is not satisfactory and the cause is doubtful, as in Cases 1, 2 and 3, it is advisable at first to culture the sputum for resistant organisms and then to test for systemic diseases such as fibrocystic disease of the pancreas (mucoviscoidosis) or agammaglobulinaemia. Pittman (1960) tested 52 patients with established bronchiectasis for gammaglobulin concentration in the blood and found no deficiency, but she reports a case of bronchiectasis with severe hypogammaglobulinaemia. Other cases have been reported in the literature.

The prognosis in bronchiectatic cases with asthma is never good. Even if all bronchiectatic areas are removed, the symptoms persist. Strang (1956) states 'the presence of asthmatic symptoms before operation does seem to influence the result'. In patients treated conservatively it may add to the pulmonary strain thus producing emphysema, pulmonary hypertension and cardiac failure as in Cases 5, 6 and 7 (Table 19).

In the present series there were 38 cases in the medically treated group in which it was impossible to say whether the tubular dilatation was likely to be permanent. In a few it was permanent as proved by repeat bronchography and some of these have been subjected to lobectomy; in others the symptoms have persisted, suggesting a permanent lesion, but in many all symptoms and signs have disappeared, and it is possible that in some of these the tubular dilatation has reverted to normal (Fig. 14a and b). This reversible phenomenon in children is important to remember, otherwise unnecessary lobectomy

may be performed. Williams and O'Reilly (1959) describe two pathological types of bronchiectasis: (*a*) that with subacute pyogenic pulmonary collapse and (*b*) that with non-specific infective bronchiolitis and/or interstitial pneumonia. They believe that with chemotherapy, postural coughing and physiotherapy many cases of type (*a*) will show complete or partial resolution of collapse and bronchiectasis, but that such treatment is less effective in type (*b*). It is therefore important to recognize those cases which may still be considered reversible (see later).

Surgery for bronchiectasis must of necessity be selective, but practice has changed over the years from the removal of grossly diseased lobes or lungs only, to segmental resection in an attempt to remove all diseased parts and at the present day to the more conservative surgery of removing only grossly diseased parts or definite localized bronchiectasis. The present series of cases fell largely into the middle group and many patients underwent several operations for partial resection. If patients treated medically and those treated surgically can be compared, it is that more surgically treated patients have improved whereas more medically treated patients have remained the same (Fig. 5); in Strang's series (1956) this was the case. However, if surgical selection is correct the patient ought to improve. Nevertheless, many surgically treated patients can become symptomless even when some residual bronchiectasis persists. Rosemond, Burnett and Humphrey-Long (1951) report on the follow-up of 159 surgically treated patients. In 55 patients there was residual disease yet 14 of these were symptomless. Nevertheless, the reverse phenomenon is not uncommon where post-operative bronchograms show no bronchiectasis yet the child remains with a persistent cough and symptoms. Strang (1956) remarks on this and suggests that it might be caused by sinusitis or a generalized bronchitis. If Williams and O'Reilly (1959) are correct in their pathological differentiation then type (*b*) (see above) in which there is a non-specific infective bronchiolitis and/or interstitial pneumonia will lead to a more generalized disease with persistent symptoms after the bronchiectatic lesions have been removed. Allergy may also be the cause in some patients.

In the selection of cases for surgery therefore, many important points must be considered and each patient is an individual problem. The following points may be of value:

(1) Grossly diseased parts, particularly if saccular, are better removed provided that enough healthy lung can be left.

(2) Moderately severe dilated bronchi with symptoms, if localized, are probably best removed.

(3) Cylindrical bronchiectasis with a bulbous or saccular periphery (Fig. 15b) where symptoms are troublesome is probably best removed, if it is localized, as this type is irreversible.

(4) Cylindrical bronchiectasis in children should be left alone and watched even if it is associated with symptoms. Some of it will be reversible, and some will revert to the fusiform or varicose types with a good prognosis.

(5) Diffuse bronchiectasis should be left alone unless a grossly diseased localized area is also present (see (*a*) above).

(6) If the child has asthmatic symptoms the prognosis is less promising. These patients tend to remain with their asthmatic symptoms or chronic bronchitis.

Does bronchiectasis spread? This is not easy to answer because superadded pneumonic processes or pulmonary collapse may themselves cause a spread of the disease. However, in the severe type of bronchiectasis with much sputum it is possible for adjacent dependent bronchi to be subject to tip over secretions with mild bronchiectasis. Furthermore, if the remaining lung is not completely healthy after lobectomy, the bronchi which become dependent may develop bronchiectasis or, if already dilated, may become more dilated. On the whole, bronchiectasis is not a spreading disease and usually remains confined to the lobes originally affected. Strang (1956) states 'there was no evidence from a study of these cases to support this concept of bronchiectasis as a spreading process'. But Dyggve and Gudbjerg (1958) found new dilatation in three out of 11 post-operative cases.

In the series of cases reported here no special study has been made of the part played by antibiotics in the natural history of the disease, but it is known that most (if not all) patients received antibiotics for the febrile respiratory infections from time to time. Comparison of cases occurring before the use of antibiotics and those of the present day is of little value as diagnosis of milder cases is made more frequently today, thus making the two groups incomparable. Furthermore, the old concept of the disease as a crippling disorder with a high mortality may have been related mainly to the severe types diagnosed by clubbing and fetid sputum. With the modern facilities of chemotherapy, surgery and postural drainage with physiotherapy, the history of bronchiectasis from childhood is one of considerable improvement particularly around puberty in the second decade. Whether this improvement will be maintained in adult life remains to be seen.

Summary

A clinical review of 225 cases of bronchiectasis is given; 104 of these were treated medically and 121 surgically. The patients have been followed for eight to 21 years at the time of report and their ages range from 10 to 29 years. There is a slight preponderance of females.

In 169 patients the onset of the disease was in the first five years of life; thereafter there was some improvement which became most marked in the second decade around puberty. In the twenties the condition seems to be stationary. The results were slightly better in the surgically treated patient, but the two groups are not comparable.

Of the patients, 53% have suffered from an intercurrent respiratory infection or asthma during the past six years and some have frequent attacks. None complained that the disease affected their marriage, but in one patient it created a complication.

There was no cough in 33% and in 44% no sputum. Nasal discharge, although less troublesome to some, still persisted in others and was a possible cause of cough. Wheezing at times was complained of by 42%, and undue breathlessness on exercise by 53%. This was more marked in the surgically treated patients.

Postural defects and abnormal shape of the chest were not uncommon features particularly in the surgically treated patients. A surprise finding was in the growth records for height and weight. There was a tendency for the records to be under the 50 percentile and a few patients were grossly underweight, particularly the medically treated patients.

Clubbing had disappeared in about 69% of the patients in whom it was originally reported. In 57% of the patients no moist sounds could be heard in the lungs even in patients with habitual cough.

Twelve deaths are reported since the original published records (Field, 1949): five surgically treated, five medically treated and two patients who died from causes other than the chest. All the patients except the last two were suffering from extensive bilateral disease.

It appears from the above records that the history of bronchiectasis between 1938 and 1956 in this series of cases shows an early onset mainly in the first five years of life, a gradual improvement, particularly around puberty, but with troublesome intercurrent chest infections which seemed to be well controlled with chemotherapy and antibiotics. Mortality after infancy was confined to patients with bilateral extensive disease.

I am greatly indebted to members of the Research Committee of The Hospital for Sick Children, Great Ormond Street, for allowing me to continue this follow-up study and for arranging all the necessary facilities. I am also very grateful to Professor R. S. Pilcher of University College Hospital, London, and his staff who so kindly collected the data on the surgical cases, the majority of which are being followed regularly by them, and also to Professor Pilcher for his continued advice and assistance.

I would also like to thank the staff of The Hospital for Sick Children, Great Ormond Street, and the Paediatric Department, University College Hospital, for their assistance in this follow-up study. In particular I am greatly indebted to Mr. J. B. Ready, Medical Records Officer, for his co-operation and help. Finally, I would like to thank Mr. D. Martin and the staff of the Department of Medical Illustration for the photography and charts.

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