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Bronchiectasis

Third Report on a Follow-up Study of Medical and Surgical Cases from Childhood

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Bronchiectasis is becoming a comparatively rare disease. A small investigation into the incidence of the disease in some of the children's hospitals in the United Kingdom in 1961 illustrates the period of greatest reduction in admissions (Table I). The yearly figures submitted, in most instances from 1938, showed the fall in incidence to occur mainly between the years 1952 and 1960. This was the time when the broad spectrum antibiotics, particularly the tetracyclines, became available for general use. Nevertheless at the same time surgical intervention became less popular, and this may also have affected the admission rates. In a recent review of 187 cases of bronchiectasis, Glauser, Cook, and Harris (1966) clearly showed the sudden drop in incidence of the disease as seen in the Children's Hospital Medical Center, Boston, between 1950–1956. They explain most of the reduction by the greater availability and use of antibiotics in the treatment of infection.

Bronchiectasis will remain with us as a disease but it may be more difficult in future to study its life history on this scale.

The Present Study

The two previous follow-up reports on these patients (Field, 1949 and 1961) have been mainly clinical like the present study. However, after the last report in 1961 it was felt that, compared with those of other authors, the medical group of patients included an excessive number of very mild cases who had had no symptoms since childhood. Though it would have been interesting to continue to see these patients, the difficulties were considerable so they were dropped from the survey, leaving a smaller medical group with definite moderate or severe bronchiectasis. The 104 medically-treated patients (Field, 1961) were reduced to 79 of whom 3 died, and reports on these were included in the analysis of deaths in the 1961 report (Cases 5, 6, and 7). Of the 76 patients followed, 54 were contacted in 1965, 43 of these attended for examination, and 11 replied to the questionary. Of the 121 patients treated at some time by operation, 111 were contacted, 80 of these were examined, and 31 replied to the questionary. There were no more known deaths in the medically-treated group, but 4 of those treated by operation had died.

The *observation period* since the first examination in childhood was for the medically-treated patients 17 to 25 years, with a mean of 22 years, and for the surgically-treated patients 16 to 25 years, with a mean of 21 years.

The *present ages* of the medical cases varied from 23 to 37 years, with a mean of 27·7 years, and the ages of the surgical cases fell between 19 and 37 years, with a mean of 27·5 years.

The female *sex* showed a slight preponderance with 29 of 54 medical cases and 58 of 111 surgical cases.

In order to give a general assessment of the

TABLE I

Admission Rate for Bronchiectasis per 10,000 total Admissions

	1952	1960
London		
The Hospital for Sick Children, Great Ormond Street	29	12
Liverpool		
Alder Hey Children's Hospital	37	13
Manchester		
Booth Hall Children's Hospital	49	8
*Sheffield		
The Children's Hospital	99	10
Glasgow		
The Children's Hospital	24	6

* Includes readmissions.

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TABLE II

General Assessment by Grades

Treatment	Grade I		Grade II		Grade III		Grade IV		Total	
	No.	%	No.	%	No.	%	No.	%	No.	%
Medical ..	17	31·5	21	38·9	15	27·8	1	1·8	54	100
Surgical ..	37	33·3	51	45·9	19	17·2	4	3·6	111	100
Total	54	32·7	72	43·7	34	20·6	5	3·0	165	100

patients the following system of Grades, established by Professor Pilcher, was applied to each case.

Grade I: Patients who have no habitual respiratory symptoms and lead normal lives.

Grade II: Patients who lead normal lives in spite of having habitual cough with or without nasal catarrh.

Grade III: Patients who for most of the year can lead a normal life but who have occasional bouts of respiratory illness which necessitate them staying away from work or staying in bed.

Grade IV: Patients who are more or less permanently ill or do not have sufficiently long periods of good health to hold down a job.

This report has separated the medically-treated and surgically-treated cases, and considered them under the 4 grades just mentioned (Table II).

It is interesting to note that the proportion of cases in each grade is somewhat similar in spite of the fact that there was selection for those cases treated surgically. This could suggest that bronchiectasis is, in the majority of cases, a disease process with a life history not greatly affected by surgery. It is interesting to observe that 32·7% of patients have no habitual respiratory symptoms and lead normal lives. What factors, if any, influence this favourable prognosis?

TABLE III

Patients with Bronchiectasis Caused by Tuberculosis According to Present Grade of Assessment

Treatment	Grade of Assessment		
	I	II or III	IV
Medical	4	0	0
Surgical	4	2	0

Aetiological factors. The possible aetiological factors recorded were pneumonia, whooping cough, measles, asthma, miscellaneous causes, and none, or a combination of these. No relation between aetiology and prognosis seemed to exist except in the miscellaneous group. No patient in this series gave a history of foreign body as a possible cause, but in those whose bronchiectasis was probably the result of or related to tuberculosis, good progress had been made, and many were free from symptoms. Table III shows that patients whose bronchiectasis was caused by tuberculosis fell in significantly better grades ($p < 0·01$) than those where it was due to other causes. From this it appears that patients with bronchiectasis resulting from tuberculous infection usually have a good prognosis clinically (Fig. 1). On nearly all the patients a tuberculin (Mantoux) test was performed when first seen in childhood. Table IV gives the number strongly positive at that time and their present grade of general assessment. If the cases of bronchiectasis definitely caused by tuberculosis are excluded in Table IV, then little significance can be attached to a strongly positive Mantoux where other causes for the bronchiectasis exist. The distribution of grades among these patients does not differ significantly from that in the remainder of the series.

Extent of disease. As would be expected, the outlook is less good for patients with bilateral disease. Table V shows that patients with bilateral

TABLE IV

Patients with Strongly Positive Mantoux when First Examined According to Present Grade of Assessment

Treatment	Grade of Assessment			
	I	II or III	IV	Died
Medical	6	7	0	1
Surgical	7	5	0	0

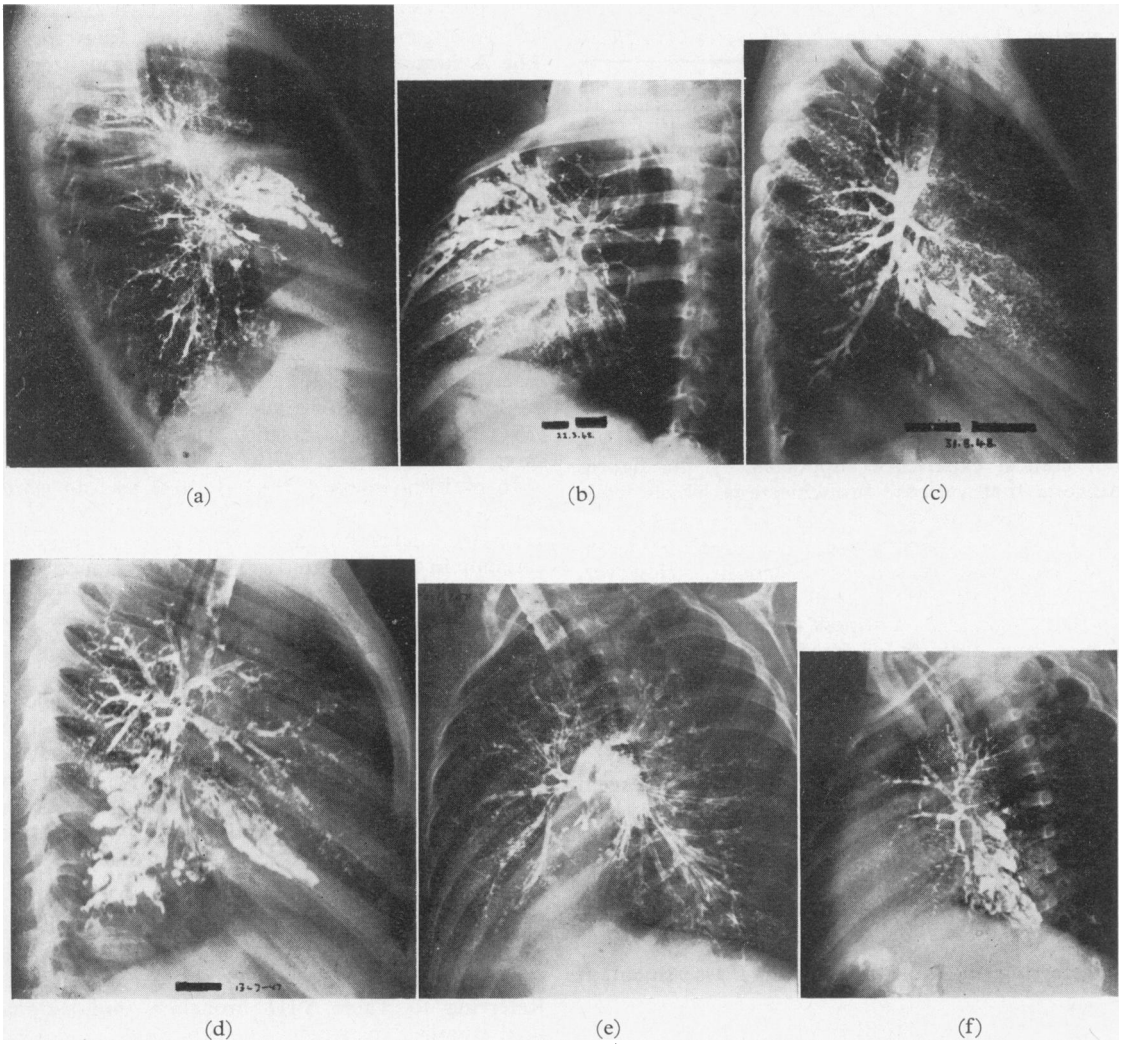

FIG. 1.—*Bronchograms of patients suffering from tuberculosis and bronchiectasis. All now assessed as grade I except Case 9 assessed as grade II.*

(a) *Case 5. Long illness with extensive tuberculous disease of right lung. Acid fast bacillus isolated. Calcification. No surgery.*

(b) *Case 6. Long history of tuberculosis in infancy with calcification in left upper zone. Bronchiectasis left upper lobe including lingula removed surgically.*

(c) *Case 7. Treated for tuberculosis as a child in a sanatorium. Father died of tuberculosis. Calcified hilar glands in* x-*ray of chest. Bronchiectasis right middle and part of lower lobe. Right middle lobectomy only.*

(d) *Case 8. Pneumonia 9 months previously which failed to resolve. Tuberculin test 1 in 10,000 strongly positive at 4½ years of age. Bronchiectasis right middle and lower lobes with a filling defect seen in the postero-anterior bronchogram. Surgical removal of right middle and lower lobes. Extensive matting together of glands suspected to be tuberculous.*
(e) and (f) *Case 9. Complete obstruction of left lower bronchus by hilar gland seen at bronchoscopy and in the bronchogram* (e). *Tuberculin test strongly positive at 4½ years of age. The bronchogram (f) taken 1 year later shows cystic dilatation of the left lower and part lingula bronchi. Surgical removal of the left lower lobe and part lingula 5 years later.*

TABLE V

Extent of Disease According to Grade of Assessment

Treatment		Grade of Assessment			
		I	II	III	IV
Medical	Unilateral	12	7	3	1
	Bilateral	5	14	12	0
Surgical	Unilateral	22	18	7	1
	Bilateral	15	33	12	3

The figures denote number of patients.

disease fell in significantly poorer grades ($p < 0{\cdot}01$) than those with unilateral disease.

Type of bronchiectasis. It would be unwise to draw too many conclusions from Table VI, but clinical experience supported by the figures suggests that varicose bronchiectasis which tends to be diffuse usually has a good prognosis, and saccular bronchiectasis alone, successfully removed by operation, also has a good prognosis. However, tubular bronchiectasis tends to be diffuse and related to a general 'chestiness' and often wheeziness, with a poor outlook for freedom from symptoms. The figures for fusiform bronchiectasis are small and indecisive, but a better prognosis had been anticipated.

Age at onset of disease. This did not seem to be related to the prognosis. In this series most patients acquired the disease at or under 5 years of age: 45 medical cases (83·3%) and 96 surgical cases (86·5%) fell into this group.

Symptoms

Referring to Table VII, grade I assessment, it will be noted that 15 (88·2%) medical cases and 22 (59·5%) surgical cases were free from *cough*. The 4 surgical cases with persistent cough were assessed grade I because the persistence of the cough was not thought to be due to the bronchiectasis but rather to disease of the upper respiratory tract or to smoking. There were 14 (82·4%) *sputum*-free medical patients in grade I and 26 (70·3%) surgical patients. It should be noted that practically all patients in grades II, III, and IV had some cough and some sputum. This is to be expected according to the definition.

TABLE VI

Type of Bronchiectasis According to Grade of Assessment

Type of Bronchiectasis	Treatment	Grade of Assessment			
		I	II	III	IV
Varicose only	Medical	5	0	1	1
	Surgical	3	3	1	0
Saccular only	Medical	1	1	1	0
	Surgical	13	7	4	1
Tubular only	Medical	4	11	5	0
	Surgical	9	20	2	1
Fusiform only	Medical	1	3	2	0
	Surgical	3	3	2	0

The figures denote number of patients.

Haemoptysis. This was absent in all medical cases assessed as grade I and in 33 (89·2%) surgical cases, but there was a moderate incidence in other grades. The total for grades II, III, and IV was 13 (35·1%) medical cases and 28 (37·8%) surgical cases.

In patients assessed grade I there was no *nasal discharge* in 14 (82·4%) medical and 20 (54·1%) surgical patients but the condition was relatively common in the other grades. As already mentioned symptoms of cough and sputum can be caused by sinusitis alone. Daily *wheezing* was relatively uncommon. Not infrequently it indicated a poor prognosis. 3 out of the 5 grade IV patients had a daily wheeze. Not all patients assessed grade I were free from the occasional wheeze. The symptom of *breathlessness* is not of much value, being more a subjective feeling rather than an indication of lung function, and often depends on the normal amount of exercise taken. One patient said he was breathless on excercise and at rest yet he was finally assessed grade I.

General health for past 9 years. Of importance in assessing the present health of the patient was the history of the past 9 years. Referring to Table VIII bronchitis and asthma in the grade I patients was infrequent, but in the other grades it tended to be more frequent and debilitating. Chest illnesses were a feature of the definition of grade III patients. It should be stated here that 6 patients in grade II had had an operation on their chest during the past 9 years.

Marriage and children. Table IX gives the percentage of patients who have married. Two women complained of infertility and one was sterilized because of her chest condition after having two children. Several women complained of increase of symptoms during pregnancy including cough, haemoptysis, or bronchitis. Of 121 babies born to these patients, 4 had 'chest trouble'. On the whole, marriage seems to make little difference to the progress of the disease, though possibly

TABLE VII

Symptoms Recorded According to Grade of Assessment

Treatment	Medical				Surgical			
Grade of assessment	I	II	III	IV	I	II	III	IV
Total no. of patients	17	21	15	1	37	51	19	4
Symptoms								
Cough:								
None	15	0	1	0	22	0	3	0
Slight	2	18	9	1	14	48	10	0
Severe	0	3	5	0	0	3	6	4
No record	0	0	0	0	1	0	0	0
Nature: Persistent	0	16	10	1	4	41	15	4
Nature: Intermittent	2	5	4	0	10	10	1	0
Sputum:								
None	14	0	2	0	26	4	2	0
Little	3	8	6	1	11	36	7	0
Egg-cupful	0	9	3	0	0	9	5	1
Much	0	4	4	0	0	2	5	3
Haemoptysis:								
None	17	11	12	1	33	35	8	2
Frequent	0	2	1	0	0	1	2	0
Occasional	0	8	2	0	3	14	9	2
Nasal discharge:								
None	14	5	2	0	20	12	2	0
Little	2	8	11	0	12	29	10	3
Much	1	7	2	1	2	10	6	1
No record	0	1	0	0	3	0	1	0
Wheeze:								
None	14	8	2	0	25	21	1	1
Occasional	3	10	12	0	11	26	14	1
Daily	0	2	0	1	1	3	4	2
No record	0	1	1	0	0	1	0	0
Breathlessness:								
None	10	8	2	0	29	27	3	0
On exercise	5	13	13	1	8	22	16	1
Also at rest	1	0	0	0	0	2	0	3
No record	1	0	0	0	0	0	0	0

Figures indicate number of patients.

TABLE VIII

Illnesses Experienced over Past 9 Years According to Grade of Assessment

Grade	Treatment	Illnesses		
		Pneumonia	Bronchitis	Asthma
I	Medical	0	4 (23·5%)	0
	Surgical	0	11 (29·7%)	1
II	Medical	4 (19·0%)	13 (61·9%)	1 (4·8%)
	Surgical	6 (11·7%)	26 (51·0%)	6 (11·7%)
III	Medical	3 (20·0%)	14 (93·3%)	1 (6·7%)
	Surgical	6 (31·6%)	19 (100%)	5 (21·1%)
IV	Medical	0	1 (100%)	0
	Surgical	3 (75·0%)	4 (100%)	1 (25·0%)

* Figures indicate number of patients. Percentages relate to totals for each grade, medical and surgical separately.

TABLE IX

Number of Patients who have Married

Treatment	Marriage State	Grade of Assessment			
		I	II	III	IV
Medical	Single	5 (29·4%)	7 (33·3%)	2 (13·3%)	1 (100%)
	Married	12 (70·6%)	14 (66·7%)	12 (80·0%)	0
				1 no record	
Surgical	Single	9 (24·3%)	13 (25·5%)	7 (36·8%)	2 (50%)
	Married	28 (75·7%)	38 (74·5%)	11 (57·9%)	2 (50%)
				1 no record	

Percentages relate to grades of assessment.

separation from an over-solicitous parent may give an impression of improvement.

General health. Each patient was asked to state whether over the past 9 years he felt better, worse, or about the same. Table X records the answers. More patients treated surgically had improved as might be expected following earlier operations. Patients assessed grade IV were all either worse or the same. The total number of patients who said they felt worse is small 5·6% medical and 7·2% surgical. This subjective answer is of doubtful value except perhaps as an indication of the mental attitude of the patient to his illness.

Physical signs. Examinations were carried out on 43 patients treated medically and 80 treated surgically. Table XI gives the results. *Posture* was most commonly defective in those assessed grade III and IV where repeated respiratory tract infection was experienced. Curvature of the spine or flattened chest was most prevalent in the surgical patients. *Chest expansion*, however, was best in the surgical patients, perhaps because breathing exercises were stressed in the pre- and post-operative period. The exception to this was among the patients in grade III and IV. *Emphysema* was rarely obvious but again was most often suspected in those assessed grade III and IV. The absence of *clubbing* is significant. Only 8 cases in all (6·5%) showed evidence of clubbing, whereas in the original 160 cases recorded by Field (1949) 43·7% showed clubbing. This suggests that with the present use of antibiotics and to a certain extent postural drainage (though many were not practising this) sepsis has been controlled and clubbing has disappeared. Even in comparison with the 1956 survey (Field, 1961) where 12·8% showed clubbing, the trend is still disappearance of clubbing. Death of the severe cases may, of course, affect these figures. Clinical evidence of *nasal secretion* compares favourably with the subjective symptoms as recorded in Table VIII. In patients assessed grade I, 8 (67%) medical and 14 (58%) surgical patients were free from nasal secretion at the time of the examination. However, in two-thirds of all the cases there was clinical evidence of upper respiratory tract infection. Referring to the original report on these patients (Field, 1949) and the 1956 survey (Field, 1961) the

TABLE X

General Health of Patient over Past 9 Years According to Grade of Assessment

Treatment	General Health During Past 9 Years	Grade of Assessment				Total
		I	II	III	IV	
Medical	Same	10 (59%)	9 (43%)	7 (47%)	0	26 (48·1%)
	Improved	7 (41%)	11 (52%)	7 (47%)	0	25 (46·3%)
	Worse	0	1 (5%)	1 (6%)	1 (100%)	3 (5·6%)
Surgical	Same	14 (38%)	14 (27%)	4 (21%)	2 (50%)	34 (30·6%)
	Improved	23 (62%)	35 (69%)	10 (53%)	0	68 (61·3%)
	Worse	0	2 (4%)	4 (21%)	2 (50%)	8 (7·2%)
	No record	—	—	1	—	1 (0·9%)

Figures indicate number of patients. Percentages relate to grades.

TABLE XI

Physical Signs Recorded According to Grade of Assessment

Treatment	Medical				Surgical			
Grade of assessment	I	II	III	IV	I	II	III	IV
No. of patients examined	12	18	13	0	24	36	17	3
Posture								
Good	7	8	5	—	22	21	4	0
Slight defect	5	8	6	—	2	11	8	2
Bad	0	1	1	—	0	4	5	1
No record	0	1	1	—	0	0	0	0
Shape of chest								
Normal	8	13	10	—	14	16	7	0
Pigeon or barrel	0	2	3	—	0	2	0	1
Flattened	2	2	1	—	8	14	6	2
Curvature of spine	2	3	1	—	2	8	7	1
Chest expansion								
Good	1	3	3	—	10	18	3	0
Average	7	12	7	—	12	12	4	1
Poor	4	3	3	—	2	6	10	2
Emphysema								
Obvious	0	0	1	—	0	0	0	2
Probable	0	6	4	—	4	12	10	1
Clubbing								
Absent	12	17	12	—	24	35	13	2
Present	0	1	1	—	0	1	4	1
Nasal secretion								
None	8 (67%)	3	2	—	14 (58%)	7	4	0
Slight	3	7	7	—	9	21	7	1
Moderate or blocked	1	8	4	—	1	8	6	2
Moist sounds								
None	8	6	2	—	23	22	6	1
Few	4	9	7	—	1	14	7	0
Many	0	3	4	—	0	0	4	2
Localized	3	4	3	—	0	9	2	0
Diffuse	1	8	8	—	1	5	9	2
Prolonged expiration		3	2	—	0	1	0	1

Figures denote number of patients.

improvement which frequently occurred at puberty has not been maintained. The absence of *moist sounds in the lung* (Table XII) was more commonly experienced in the surgical (65%) than in the medical patients (37·2%). This same difference can also be observed in the 1956 survey (Field, 1961). Understandably, removal of the diseased part should reduce or eliminate the infected area. There appears to be deterioration of the medical patients in the present survey, fewer being free from moist

TABLE XII

Comparison of Moist Sounds in Lung Since Original Survey

Moist Sounds in Lungs	No. of Patients				
	When First Seen (Field, 1949)	1956 Survey	(Field, 1961)	Present Survey 1965	
		Medical	Surgical	Medical	Surgical
None	16	55 (55%)	74 (63·2%)	16 (37·2%)	52 (65%)
Few	93	29	39	20	22
Many	116	15	4	7	6
No record	0	5	4	0	0

TABLE XIII

Analysis of Deaths

Case No.	Age at Death (yr.)	Sex	Extent of Bronchiectasis	Pulmonary Resection	Cause of Death	Necropsy
1	25	M	Gross cystic left lung; some fusiform right middle and lower lobe with crowding of bronchi	Left pneumonectomy	Died in cardiac failure with cor pulmonale; right upper and middle lobes markedly emphysematous with gross congestion and oedema; right lower lobe small with great dilatation of bronchioles	Yes
2	28	F	Right middle and lower lobes; left lower and lingula lobes (varicose-tubular)	Right middle and lower lobes; left lower and lingula lobes	Pneumonia both remaining upper lobes	Yes
3	29	M	Isolated cysts right middle and part right upper lobe; ? congenital (asthmatic patient); later diffuse bronchiectasis both lungs	Right middle and part of right upper lobe	Cardiac failure; patient was a bad bronchitic and asthmatic	No
4	31	M	Diffuse right lung; left lower and lingula (asthmatic patient) (sister has bronchiectasis)	Left lower lobe performed in Canada	Congestive cardiac failure probably cor pulmonale (from parents' report)	No

sounds, but this may be because 25 of the mildest cases in the 1956 survey have been dropped from the present survey.

Analysis of fatal cases. Three patients treated medically died shortly after the last survey in 1956 and were therefore included in the analysis of deaths in that report (Field, 1961). Since then four patients, all treated at some stage with operation, have died (see Table XIII).

It is remarkable that Case 1 lived as long as 25 years. When first seen at the age of 5 years he was a bad bronchiectatic, with gross clubbing and much sputum. He improved on medical treatment so his grossly cystic left lung was removed in spite of disease on the right side (Fig. 2a, b, and c). Three years after the pneumonectomy a bronchogram showed bronchiectasis, with crowding of the bronchi in the right middle and lower lobes and emphysematous expansion in the right upper lobe herniating over to the left side of the chest. A repeat bronchogram three years later showed cystic dilatation of the bronchi in the right middle and lower lobes. The child himself improved to a certain extent but spent a good deal of his life in convalescent homes, special schools, or hospitals. The question here is whether it was justifiable to remove the grossly diseased part knowing the other lung was diseased. It probably prolonged life and improved his symptoms, particularly the volume of sputum, to a certain extent.

Case 2 had repeated attacks of pneumonia in infancy and had been 'chesty' most of her life. It is now generally appreciated that such patients are poor subjects for operation, as their disease is a generalized one.

Case 3 is important as his original disease seemed to consist of isolated cystic lesions, probably of congenital origin, but he was an asthmatic (Fig. 2d). Though the cystic lesions were successfully removed the whole bronchial tree seemed to be weak, as a bronchogram 7 years later showed diffuse bronchiectasis. There is no doubt that asthmatics are poor subjects for operation.

Case 4 was treated medically for many years because of the diffuse nature of his disease. At the age of 19 years, however, when he was in Canada, the grossly diseased left lower lobe was removed. It is doubtful if this made very much difference to the progress of his disease. Temperamentally he was difficult and at times uncooperative. His sister also suffers from generalized (fusiform) bronchiectasis, but her general health is reasonably good (grade II). Tests for fibrocystic disease of the pancreas have not been done.

Discussion

This further report on the long-term study of cases of bronchiectasis from childhood shows that

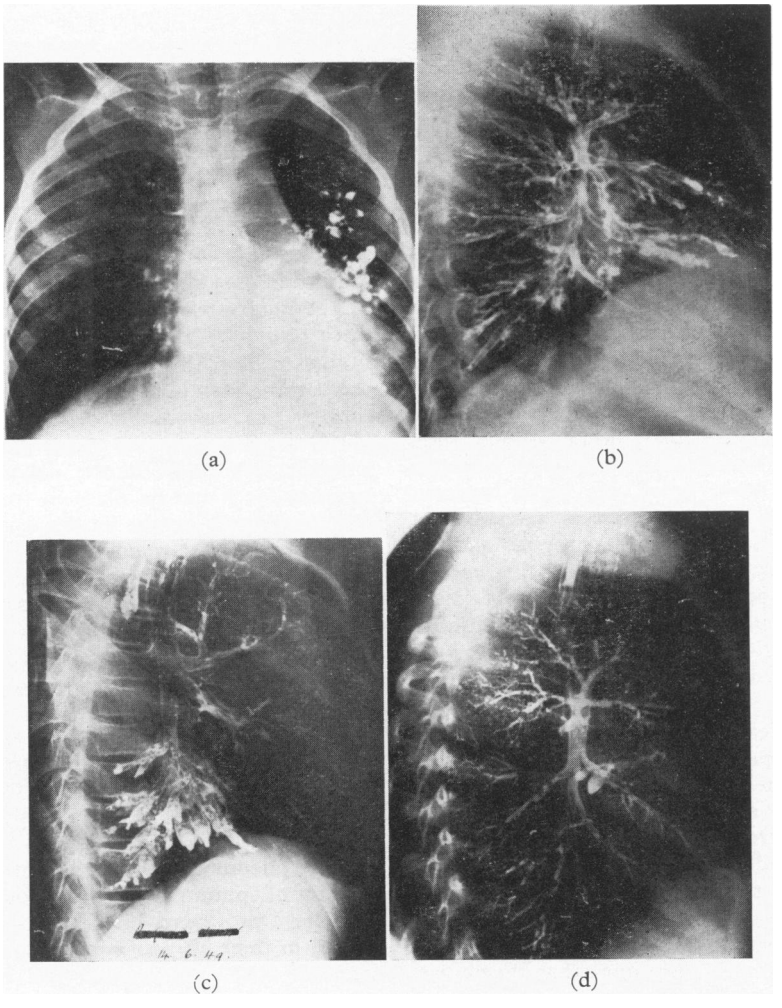

FIG. 2.—*Bronchograms of two fatal cases: (a)-(c) Case 1 who lived for 25 years (Table XIII); and (d) Case 3 who lived for 29 years (Table XIII).*

(a) *Postero-anterior bronchogram when patient was 5 years old showing cystic dilatations of the left lung.*

(b) *Right lateral bronchogram showing slight diffuse irregularity of the whole bronchial tree.*

(c) *Right lateral bronchogram 6 years after left pneumonectomy showing cystic dilatation of the right middle and lower lobe bronchi, with crowding. The right upper lobe has expanded and shows emphysema and herniation.*

(d) *Right lateral bronchogram showing three cysts probably congenital in origin.*

After surgical removal of right middle and part right upper lobe, diffuse bronchiectasis in the remaining lung developed. The patient was an asthmatic and lived for 29 years.

the patients can be divided into those who are consistently symptom free (grade I assessment) and those who have persistent symptoms, most commonly cough with or without sputum (grades II, III, and IV). It would be interesting to know what factors influence the ultimate progress of this disease. One aetiological factor—tuberculosis—appears to carry a good prognosis, though the numbers are small. Only those patients whose bronchiectasis was considered to be caused by tuberculosis were included in this category. All the 4 medically treated patients and 4 out of 6 surgically-treated patients were assessed grade I (symptom free). The presence of a strongly

positive tuberculin (Mantoux) test alone was of little significance where other aetiological factors existed. As recently pointed out by Rosenzweig and Stead (1966), the tuberculous process itself may be a necrotizing pulmonary infection leading to bronchiectasis without bronchial obstruction or secondary infection. It is the absence of secondary infection which probably gives the good prognosis. Diaconită and Săvuleanu (1966) discuss from a study of 81 specimens the effect of modern chemotherapy on the pathological process in tuberculous bronchiectasis. They describe a modification of the bronchiectatic lesion, leaving cylindrical dilatation but intact bronchial walls as if the process had healed over. The question of surgical intervention is a difficult one in these tuberculous patients, but as secondary infection may occur at any time, localized areas of bronchiectasis with poor drainage, particularly the lower lobes, are better removed after the active tuberculous process has subsided. Upper lobe disease tends to remain clear and uninfected providing the bronchi have not become dependent. Rzepecki, Zuk, and Zalewski (1966) record a 4- to 10-year follow-up of 55 children treated by operation. Nearly 100% recovery was achieved in patients with bronchiectasis of tuberculous origin.

It has been clearly stated in this and previous reports that the patients treated medically are not comparable with the patients treated surgically because the latter are of necessity a selected group. Nevertheless there is a surprising similarity in the follow-up studies over the years as if there is a natural history of the disease process even if all the apparent disease has been removed surgically. It is now well recognized that the initial aetiological factor may damage or change more than the observable bronchiectatic area, or may affect the patient's immune reaction to infection and thus prevent complete amelioration of symptoms even if all the disease has been removed. Avery, Riley, and Weiss (1961) suggest the possibility of a congenital predisposition to bronchiectasis in some patients. Nevertheless certain aetiological factors such as tuberculosis or foreign body may have only a local effect so that removal of the diseased part or good medical drainage of the part will result in absence of symptoms and a clinical cure. There still remains a group of patients with local, scattered, or diffuse disease whose prognosis remains unpredictable. It is clear, however, that asthma in association with bronchiectasis has an unfavourable prognosis. Many of the fatal cases have had associated asthma, including Case 3 in Table XIII (Fig. 2).

With the introduction of antibiotics and chemotherapy, not only has the incidence of bronchiectasis fallen sharply but the prognosis of established cases has probably improved. Fine and Baum (1966) report an interesting follow-up of applicants for flying training in World War II. 41 were found to be suffering from bronchiectasis and of these news of 24 was obtained. 3 had died but only one of these was thought to have died from a complication of his bronchiectasis. 20 out of 21 were working, 4 had had pulmonary resection, 7 were without symptoms, 10 had minimal symptoms, and 4 moderately severe symptoms. Comparing these results with our own they appear to be very similar, with a 30% symptomless group.

On these results it is difficult to assess the part surgery has played. It is doubtful if, as Clark (1963) suggests, surgery offers the best chance of a satisfactory result in the majority of children; nevertheless it has its place in treatment. Severe and moderately severe localized disease amenable to surgery is best removed, providing the disease process is not of a general nature, as evidenced by asthmatic or diffuse bronchitic symptoms. Time should be given for the bronchiectatic areas to be clearly defined. Diffuse bronchiectasis is best treated medically with postural drainage and antibiotics for exacerbations. The problem of removing only grossly diseased areas in a more diffuse involvement remains controversial. Case 1 in Table XIII is a good example (Fig. 2a, b, and c). This boy lived for 18 years, mainly on one right upper lobe. His condition was never very good but the pneumonectomy undoubtedly reduced the amount of sputum and toxicity for many years and may have prolonged his life. Borrie and Lichter (1965) in their 10-year survey of surgically treated patients state, 'Incomplete but worthwhile relief of symptoms can be obtained even in the presence of known bronchitis and minimal bronchiectasis in the remaining lung tissue.'

Finally consideration must be given to the trend of bronchiectasis as a disease process in the light of present-day treatment. In previous reports (Field, 1949 and 1961) it is clear that the patients as a whole improve in the second decade, during puberty, and remain stationary in the third decade. Some of them are now reaching the fourth decade without any marked change except for the few patients with severe disease who have died. Comparing the present grade I assessment with the previous report (Field, 1961) in which comparable patients were assessed as 'well', in the medically treated patients 31·5% are now grade I, whereas 33·7% were assessed 'well' 9 years previously. In the surgically-treated patients 33·3% are now

grade I, whereas 9 years ago 29·7% were 'well'. The difference is negligible as 25 of the mildest medically-treated patients were dropped from the survey.

Summary

This is the third report on 54 medically-treated and 111 surgically-treated patients followed from childhood. The observation period averages about 21 years and the present average age of the patients is 27 years.

The patients have been classified into four grades of assessment. Those in grade I are healthy and symptom free, and include 31·5% of the medically-treated and 33·3% of the surgically-treated patients.

In this series the only aetiological factor with an apparently good prognosis was tuberculosis.

Prognosis has been related to the extent of disease and the type of bronchiectasis.

Haemoptysis and recurrent chest infections were common in grades II, III, and IV.

Marriage did not seem to affect the course of the disease except during pregnancy when there was usually an exacerbation of symptoms.

Four deaths are reported, all in patients who had been treated surgically.

The trend has been for definite improvement to occur in the second decade, which on the whole has been maintained in the third decade and so far into the fourth decade. The exceptions are the severe cases which slowly deteriorate to a fatal outcome.

There was a sharp fall in incidence of cases of bronchiectasis attending children's hospitals between 1950 and 1960 at the time the broad spectrum antibiotics were introduced.

Present-day treatment with chemotherapy and antibiotics has probably affected the long-term prognosis of bronchiectasis.

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