THE PROGNOSIS OF BRONCHIECTASIS IN CHILDHOOD*

BY

ALFRED WHITE FRANKLIN

From the Children's Department, St. Bartholomew's Hospital, London

(RECEIVED FOR PUBLICATION JULY 5, 1957)

In England Ogilvie (1941), Field (1949) and Conway (1952) have collected cases of childhood bronchiectasis over the long term and on the large scale needed for worthwhile information. The gloomy prognostication, based on old clinical and necropsy studies, that the bronchiectatic child will not grow up or will be a burden to himself and his neighbours, is not generally true. Since 1926—that is, during 20 to 30 years of active bronchography—the diagnosis has depended on an *x*-ray appearance of dilatation in more or less of the bronchial tree. On this basis it seems now established that: (1) dilatation once present always persists, except where tubular widening is confined to lobes or segments temporarily collapsed (atelectatic reversible bronchiectasis); (2) under natural conditions the distribution of the affected areas does not alter; and (3) the clinical picture changes little and the tendency is for any change to be for the better even without surgery and without chemotherapy.

In 1944, I (Franklin, 1944) reviewed 24 personal cases of bronchiectasis, observed for periods of four and a half to 13 years, and found 15 of them completely well and free from symptoms, while four were assessed as invalids, and five were classed as delicate according to criteria described below. The *x*-ray evidence of bronchiectasis had remained constant in distribution throughout the period of observation in all but one of the cases. This boy had the severest form of bronchiectasis clinically, with the added complications of steatorrhoea, an anaemia of pernicious type, and for a short period signs of subacute combined degeneration of the cord. Despite all this and the fact that the bronchiectatic areas spread he is, at the age of 23, well grown, normal and healthy and has been free from respiratory symptoms for six years. With a further follow-up it is possible to take from this whole group 19 cases with an interval from the onset of symptoms of between 13 and 26 years, and one with an interval of 34 years. In all but two the period of observation exceeds 10 years. Of the 20 patients (Table 1), one died aged 20 years of cerebral abscess (before 1944) and three are invalids with severe cough and much purulent sputum. Three, despite large amounts of sputum, are usually well and in light employment. Two are well except for occasional attacks of pneumonia. The remaining 11 patients are completely well and free from symptoms. Of those graded well in 1944, in only one case has there been deterioration, with the copious production of sputum, following an A1 assessment and entry into the British Army. In all the others any change has been for the better.

In 1945 the Meath School of Recovery (for the care of bronchiectatic children aged 5 to 8 years) was opened by the Invalid Children's Aid Association of London through the stimulus of Dr. Elaine Field. I had to assess cases for admission and to see them over their six to 18 months' stay, and have recently reviewed them all or seen clinical reports or *x*-ray series from their own paediatricians or chest surgeons. Out of 191 admissions between January, 1946, and December, 1952, eight were non-proven cases, 12 were inadequately followed up and the remaining 171 are included in the survey. Children were admitted because too ill to attend day school from home; for convalescence and schooling before, between and sometimes after lung resection; for social reasons, for example, to allow an unmarried mother to work; or when home conditions were considered too bad for a child with chronic respiratory disease.

As the school provided physiotherapy and postural drainage the amount of sputum and cough was stressed at first, but it soon became evident that some ill and undernourished children had little sputum or cough, while others with a great deal of both were extremely well and full of energy. The cases were then grouped (Table 2) as invalid (38 cases), delicate (70 cases) and well (63 cases). The label *invalid* is applied where sputum is of inexhaustible amount or there is failure to

* A paper presented to the combined meeting of the American Pediatric Society, British Paediatric Association, Society for Pediatric Research and Canadian Paediatric Society, Quebec, June, 1955.

TABLE 1

LONG-TERM ASSESSMENT OF 20 PREVIOUSLY REPORTED CASES OF BRONCHIECTASIS (FRANKLIN, 1944)

Assessment in 1944 and Case No.	Age at Follow-up (years)	Observation Period (years)	Age at Onset (years)	Duration of Symptoms (years)	Final Assessment
Invalid .. 3	20	10	4	16	Died aged 20 years
4	20	12	9/12	19	Delicate (1944)
6	18	12	4	14	Invalid (1944)
7	25	19	3/12	25	Well
11	19	15	2	17	Invalid
Delicate .. 6	18	6	1	17	Delicate (1944)
12	23	16	Birth	23	Invalid. (Left lower lobectomy at age 19)
19	25	21	6/12	25	Age 23: empyema. Age 25: light employment. (Gross sputum)
21	25	22	2	23	Delicate. Light employment. (Gross sputum)
Absolutely well 1	34	22	3/52	34	Well
2	18	9	8/12	18	Well (1944)
5	27	17	6/12	26	Well
9	23	12	4	19	Well
10	23	20	Birth	23	Well, but treated for duodenal ulcer
13	20	14	4	16	Well
15	26	19	6	20	Well
17	22	10	8	14	Well
18	15	11	2	13	Well
20	22	15	4	18	Well
24	22	19	3	19	Well. (A1 in Army. Gross sputum)

thrive or crippling bronchitis or asthma; a child is labelled *well* not only in the absence of symptoms but also, if growth and energy are normal, when there are minimal amounts of sputum easily cleared, or periodic bronchitis, asthma or fever. The middle group, called *delicate*, have sputum that can be cleared though with difficulty, are capable of some exertion, but tire easily and cannot be described as well. The dividing lines, though not exact, are drawn by one observer.

At the final assessment with the same grouping standards, 15 are invalid, 23 delicate, three have died and 130 out of 171 are well, the follow-up time (not the duration of disease) being three to 10 years. Those classed as well are well grown and lead a full, normal life at school or in employment; 91 are completely free from symptoms, but 39 have traces of sputum, occasional cough, bronchitis or febrile attacks, without notable interference with the conduct of life. Table 2 shows that of 38 originally classed as invalid 14 are well, while of 63 originally well three have become invalid. All three have been operated on, none with complete removal of all affected areas. Three patients have died, one from chloramphenicol treatment, one from cor pulmonale at 16 years after a temporary improvement following partial surgery. The third died as the result of anaesthesia for bronchography, and a fourth might have joined her but for a timely tracheotomy. In these deaths treatment or diagnosis is deeply involved. This might at least discourage the routine annual stocktaking bronchogram.

Tradition demands a statement about finger-clubbing, which in this series was a measure of the degree and activity of infection, disappearing with symptomatic improvement or recovery. The more lobes involved—'lobes' meaning lobe or segment of a lobe—(Table 3) the worse the condition, and much purulent sputum is more likely with saccular than with tubular dilatation. Collapse is commonly reported in radiographs of bronchiectasis, but except in the 'well' group there is usually dilatation of

TABLE 2

FINAL ASSESSMENT OF CASES GROUPED ACCORDING TO ORIGINAL ASSESSMENT

Assessment at Entry to Meath School	Total Cases	Final Assessment			
		Invalid	Delicate	Dead	Well
Invalid	38	10	12	2	14
Delicate ..	70	2	6	1	61
Well	63	3	5	0	55
Total ..	171	15	23	3	130

TABLE 3

NUMBER OF LOBES AND TYPE OF DILATATION IN EACH GROUP

No. of Lobes	Assessment at Entry			Total (171 cases)
	Invalid (38 cases)	Delicate (70 cases)	Well (63 cases)	
1	3	19	18	40
2	6	17	13	36
3	10	18	20	48
4	15	14	11	40
5	4	2	1	7
Saccular ..	22	18	8	48
Fusiform ..	2	3	2	7
Tubular ..	14	49	53	116

bronchi outside the collapsed area and a permanent airless shadow may well be cast by a shrunken fibrous lobe.

Evidence of upper respiratory tract disease was present in all but 30 of the cases (Table 4). In another 46 cases there was either a history of removal of tonsils and adenoids or a simple nasal discharge, or both. In at least half of the invalid and the delicate cases upper respiratory symptoms contributed to the clinical picture. The relation between upper respiratory infection in general, and sinus disease in particular, and bronchiectasis is open to dispute, and the problem of which causes which may be solved by regarding bronchiectasis as sometimes part of a disease of the entire respiratory tract, improvement in one part leading to improvement in the other, but improvement in both being needed for the relief of symptoms. Lobectomy alone has accomplished little in the presence of chronic bronchitis and sinusitis, but if sinus or aural disease can be cleared up the lung condition usually improves.

TABLE 4
UPPER RESPIRATORY INFECTION

	Assessment at Entry		
	Invalid (38 cases)	Delicate (70 cases)	Well (63 cases)
Nasal discharge	19	28	23
Sinus infection	19	28	23
Sinus operation	13	21	17
Aural discharge	6	9	13
Aural operation	1	3	3
Removal of tonsils and adenoids	16	45	41
Nasal discharge or removal of tonsils and adenoids alone or in combination	8	25	13
No upper respiratory disease	12	10	8

The early onset of symptoms is striking, these starting before the age of 2 years in 99 of 171 cases (Table 5). Some prognostic importance for this is suggested by the fact that this onset of symptoms before the age of 2 years occurred in 13 of 15 judged invalid at the final assessment, in five of the seven cases where all five lobes are involved in 19 of the 20 with severe nutritional disturbance and in 20 of the 22 cases of saccular dilatation in those classed as invalid at entry. That this cannot be blamed on the character of the onset of symptoms is shown in Table 6, which compares the mode of onset in all cases and those beginning under the age of 2 years. There is no appreciable difference in the percentage distribution. Could the greater severity of the disease when the onset is early be due to infection of the lung at a particular stage of its growth?

TABLE 5
AGE AT ONSET OF RESPIRATORY SYMPTOMS

Age at Onset (years)	No. of Cases		Age at Onset (years)	No. of Cases	
Under 1	61	99	5–6	7	
1–2	38		6–7	2	15
2–3	30		7–8	2	
3–4	18	57	8–9	3	
4–5	9		Uncertain	1	

TABLE 6
CHARACTER OF ONSET

Type of Onset	Total (171 cases)	Per cent. of All Cases (100)	Onset Under 2 Years (99 cases)
From birth	6*	3·5	6
No acute illness	56	32·8	35
Pneumonia	51	29·8	31
Whooping-cough	32	18·6	16
Measles	19	11·1	9
Other	7	4·2	2

* Plus one case of neonatal pneumonia.

NOTE: Only one case presented as tuberculosis, but four more were probably cases of primary infection, and tuberculosis was present in at least 12 cases, all arising after the age of 2 years.

Although elaborate classifications of bronchiectasis have been suggested in the past, we are inclined now to think and speak of and to treat bronchiectasis as an entity. Accuracy in prognosis might be helped by classifying cases into aetiological groups and clinical types. Certainly the progressive and serious lung lesions in cases of pancreatic fibrosis should be excluded. At the other end of the scale atelectatic unilobar disease should also be excluded unless time proves that the changes persist following expansion of the lung. Empyema complicating pneumonia at onset was found in seven cases, and all these benefited from surgery. The role of tuberculosis is more difficult to determine. The tuberculin state is known only in 50 cases, and was positive in 15 and negative in 35. Only one case presented as tuberculosis, but review suggests that it was important in 11 others, four with primary lesions and seven with collapse or bronchostenosis secondary to enlarged lymph nodes. The particularly suggestive clinical features are an onset after the age of 2 years, involvement localized to one lobe or one lung, temporary finger-clubbing (in two only while there was cough and sputum), a high incidence of 'collapse' and a low one of upper respiratory disease. All cases but one are now in the 'perfectly well' group. Surgery is usually unnecessary for the relief of cough and sputum, and its use depends on the general policy adopted for the surgical treatment of childhood intrathoracic tuberculosis.

Prognosis has been considered in relation to the major clinical types of the disease. In the classical case there is much purulent sputum, often abnormal physical signs in the chest and finger-clubbing. The patient at rest punctuates time with the typical cough, a deep and effortless rattle, while exertion, emotion or changes of posture cause periodic productive paroxysms. There is a small group with some clinical symptoms and signs suggesting the disease, but with bronchitis and asthma as the prominent features, and a still smaller group with recurrent fever and occasional respiratory signs and symptoms. Haemoptysis as the sole evidence of bronchiectasis must be rare in children. Table 7

TABLE 7
ASSESSMENT AT ENTRY AND TYPE OF CLINICAL PICTURE

Assessment at Entry	Total Cases	Classical: Cough and Purulent Sputum	Bronchitic and Asthmatic	Recurrent Febrile Attacks
Invalid ..	38	36	2	0
Delicate ..	70	52	14	4
Well ..	63	41	11	11*
Total ..	171	129	27	15

* Including one case of inhaled foreign body and one of tuberculosis.

correlates the assessment at entry with these clinical types, and shows the high incidence of classical cases in the invalid group in contrast with the low incidence of invalids in either bronchitic or febrile cases. The large number of patients, 40, who are classed as well in the group labelled classical needs comment. These children had a classical history and many had the clinical evidence of bronchiectasis at the preliminary examination and even on arrival at the home. The picture though clear was always mild and was short-lived, so that within a few days or weeks of arrival neither cough nor expectoration could be induced by exercise, tipping or tapping.

This review covers the most active period in the surgical attack on children with bronchiectasis. A total of 71 cases were operated on, including 60 of the 129 classical cases, 26 of which had complete excision of all affected areas and 34 excision of part only. Some of these operations, as judged by the clinical rather than the radiological picture, seem to have been cosmetic, and some to have been carried out to allay the unjustified fear of spread of disease or of deterioration of health. Where there was complete resection of all areas known to be affected 17 of 26 patients are well, but nine retain symptoms of cough and sputum. Of the 34 with partial surgical treatment, 12 are well despite the continued presence of bronchiectatic areas. In one-half of the non-surgical classical cases the patients are well. This group contains milder cases, but also 14 patients with large amounts of sputum who escaped surgery because of the widespread disease. Of the 28 bronchitic cases, seven had complete, three partial and 18 no operation. Few if any would have been treated surgically now. One case of recurrent fever out of 15 had complete surgery with improvement. The presentation of figures is difficult and may be misleading, since numbers are small and anything approaching matched controls is impossible. The series lacks consistency because many physicians and surgeons using different criteria were in charge of the cases. The results may at least be claimed to show that surgery is not the complete therapeutic answer. The risk of post-operative pulmonary collapse and consequent spread of bronchiectasis is less than was feared at first, but extensive surgery may increase dyspnoea on exertion and diminish mixing efficiency in the lungs.

The prognosis of any chronic or disabling disease cannot be made on a purely anatomical basis. Sixteen patients (almost one in 10) were of sufficiently low intelligence to require special educational methods, and such cases do badly. The doctor's advice, as well as his methods of diagnosis and treatment, may affect the situation in unintended ways. The only unmanageable boy had been away from home continuously from the age of 10 months to 5 years because of respiratory illness. Twenty-nine other patients were separated from home in early life for long periods on medical advice, though only four of these were led by behaviour disturbance to the Child Guidance Clinic. In six cases great fear of the disease had been induced in parents and children by the doctor's terrible prognosis or by his arguments in favour of surgery. Repeated bronchograms, radiographs and surgical operations take a toll of morale. The record is held by one boy who had 56 x-ray exposures in two years. Thirty-five homes were judged physically unsatisfactory, and 10 children from slums undoubtedly improved after rehousing in a new area. Although in three cases operative treatment was considered wisest because of relapse on return to bad housing conditions, in the whole series the physical environment affected the results surprisingly little.

To sketch briefly the course of the commonest, the classical case, the onset is early in life following pneumonia, perhaps complicating whooping-cough or measles or both in succession. The cough, the sputum, with clubbing, cyanosis, abnormal physical

signs in the chest and chest deformities, are severe from the outset and continue for five or six years punctuated by recurrent pneumonia. In the lucky case the symptoms gradually improve and at 12 or 13 years of age the child is in comparatively good health except for some productive cough, increasing with any respiratory infection. Later in adolescence the symptoms disappear. In the unlucky case with severe and extensive bilateral saccular disease, chronic pneumonitis leads to permanent invalidism, possibly with cor pulmonale. Such cases are usually too extensive for surgery, while the use of antibiotics, despite a reduction in sputum and a disappearance of bacterial infection, leaves the secretion of copious mucus and the abnormal physical and radiological signs and causes little improvement in the patient's symptoms. By contrast, the mild case dries up with a course of the appropriate antibiotic and may remain free from symptoms for many months or years.

The results in the series reviewed seem quite good. The bad cases, the 15 who remain invalids, are bad indeed, and to prevent such cases seems the only solution. May I suggest that the prognosis is worst with an onset under 2 years of age, that the respiratory tract is often involved as a whole, the bronchial dilatation being only a part of what is wrong; that the extent of the disease is unlikely to change, so that repeated bronchography with its slight though definite risk is unnecessary; that selection of cases for operation requires prolonged observation and deep knowledge of the individual case; and that in assessing prognosis before deciding on treatment the natural history of bronchiectasis be remembered and the whole child and his environment be considered, so that treatment is avoided that might be more harmful than the disease itself.

I acknowledge gratefully the help of Miss Gosling, the Matron at the Meath Home at the relevant period, the staff of the Invalid Children's Aid Association, and the many colleagues in the fields of paediatrics, thoracic surgery and radiology, who have so unselfishly produced clinical records and radiographs and have sanctioned the follow-up visits of their patients. I would also thank the children and their parents for their cooperation, including much travelling.

References

Conway, D. J. (1952). M.D. Thesis, Cambridge University.
Field, C. E. (1949). *Pediatrics*, **4**, 21, 231, 355.
Franklin, A. W. (1944). *Proc. roy. Soc. Med.*, **37**, 576.
Ogilvie, A. G. (1941). *Arch. intern. Med.*, **68**, 395.