certainly made by complement fixation tests, but in the vast majority of cases the fever will have gone before this is done. The case is different when one is confronted by an outbreak instead of a sporadic case. The occurrence of many cases of any fever acquires an importance even when the illness is slight. As has been pointed out, the explosive epidemic of Q fever is more diagnostic than the individual case history and this is an occasion when Q might well be borne in mind. Another reason for making a diagnosis is that it indicates appropriate treatment. Now chloramphenicol and aureomycin are specific for Q, and are, of course, effective against many other micro-organisms. So much so that it is common practice to give them in any undiagnosed fever that does not react to sulphonamides and penicillin. If this be so, the sporadic case of Q may well be cured without the necessity of even thinking of the diagnosis. Lastly, one may make a diagnosis to clear the air of what the patient has not got. It is here that

chronic Q fever might be important. Continued and especially recurrent bouts of fever and chronic illness are most distressing and puzzling things. Any physician who sees them would gladly accept a new diagnosis to supplement his already exhausted stock-in-trade. It is easy enough by complement fixation to exclude the disease, but at the moment it can only be regarded as a very remote possibility.

Note

Complement fixation tests can be done by any virus reference laboratory. They are, in fact, usually done on sera sent there from a febrile patient.

Full bibliography and review of the subject can be found in:

HUEBNER, R. J., JELLISON, W. L., and BECK, M. D. (1949), *Ann. intern. Med.*, **30**, 495.

BRONCHIECTASIS IN CHILDHOOD

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Definition and Aetiology

The association of a chronic productive cough with dilated and inflamed bronchi was observed by Laennec in 1819. The first case he used to illustrate his description of this disease was a boy aged 3½ years who had copious expectoration of foetid sputum since whooping cough three months earlier and the second was a woman of 72 years who for upwards of 50 years had suffered from an habitual cough producing opaque yellow sputum, frequent haemoptyses and oppressed breathing.

Though Laennec's description was under the heading of 'Dilatation of the Bronchi,' he referred to a clinical condition which could be diagnosed during life. Until the advent of bronchography the term bronchiectasis was used for this recognizable clinical syndrome, but since then the dilemma has arisen as to whether it is justifiable to use it to signify mere dilatation of the bronchi without the clinical accompaniments. Clearly a

chronic productive cough and abnormal signs in the chest without bronchial dilatation cannot constitute bronchiectasis, but if the term is used to cover the slight temporary bronchial dilatations so often observed during bronchography then another name will have to be coined to distinguish the chronic disease syndrome from the single physical sign.

In practice, clinicians retain the connotation of bronchiectasis in the original sense of a chronic, frequently debilitating disease in which there is, besides dilatation of bronchi, a constant or recurrent productive cough, abnormal physical signs in the chest and a liability to complications such as haemoptysis, pneumonia, pyaemic abscess or suppurative cachexia which may shorten the duration of life. The symptoms and signs of this disease result from accumulation and stagnation of secretions in dilated bronchi and, though they may resolve when secretions are no longer retained, there is always a tendency towards recurrence so

long as the bronchi remain deformed and function inadequately. It is therefore the permanence or chronicity of the bronchial dilatation which is the principal feature of bronchiectasis and it has now become customary for radiologists to avoid using the term where it is judged that the dilatation is of a temporary nature.

Bronchi are dynamic structures which constrict and widen during respiration. Inflammation, an alteration in surrounding parenchyma, distension by retained secretions or actual ulceration and destruction of the walls can enlarge their calibre. Such conditions may result from many causes and the dilatation persist for a variable length of time. It can sometimes be assessed from the nature of the original cause and the bronchographic appearance that return to a normal calibre and functional capacity is unlikely ever to occur. In other cases, progress towards restitution can only be judged by prolonged observation, response to treatment and repeated bronchograms. However, it can reasonably be assumed that a bronchus which has been continuously dilated for more than two years and possibly as little as one year will not return to its former state and function. Bronchiectasis constitutes a clinical syndrome which supervenes as a complication of the original cause and owes its separate consideration both to the fact that its features usually persist after the initial cause has ceased to operate and, as with empyema, its magnitude of effect on the patient warrants definition as a disease in its own right.

Bronchiectasis is an uncommon disease which is found in about 3 per cent. of autopsies, but is only diagnosed during life in approximately 0.3 per cent. of out-patients in general hospitals. The onset of the disease is usually attributed to some illness in the past, but close enquiry often reveals an unusual liability to chest troubles from an earlier age.

It has long been known that bronchial obstruction by a foreign body, growth or enlarged tuberculous lymph glands can produce bronchiectasis. Experimental occlusion or constriction of a bronchus in animals has been shown to result in bronchiectasis within a matter of a few days or weeks and the speed of development is usually greatest when infection has been induced beyond the block. These clinical and experimental observations suggest that bronchiectasis is usually caused by some such mechanism, though with the passage of time evidence of the initial bronchial occlusion may have resolved or been destroyed. The obstructing material may have been relatively uninfected mucus in association with a pulmonary collapse or asthma, or heavily infected mucus resulting from bronchopneumonia or inhaled from the nasal sinuses. Local inflammatory swelling

of the mucosa can obstruct a bronchus and it is probable that secretions can also be retained due to a functional paralysis of the bronchial wall. Infection of secretions beyond a bronchial block leads to distension of the peripheral tubes by the pressure of accumulating exudate and ulcerative destruction of mural structures. Whatever the precipitating illness may have been, the usual processes leading to bronchiectasis are therefore bronchial obstruction and inflammation. Histological examination shows that the latter often resembles an endo-bronchial abscess in its severity. Experimentally, ligating one of the pulmonary arteries in an animal favours the development of bronchiectasis in that lung. It may prove that a vital step in the chain of events leading to bronchiectasis is thrombosis of bronchial or pulmonary blood vessels.

It has been suggested that reduction in volume of lung parenchyma produces such alterations in the intrathoracic pressure equilibria that the bronchi undergo secondary dilatation. Such effects are, however, very temporary and soon compensated by over-expansion of other normal lobes and shift of the mediastinum. Peribronchial and parenchymal fibrosis have been credited with the ability to widen bronchi, but the former is more likely to constrict them and before the latter can exert a traction force it must have an external purchase. Adhesions about a bronchiectatic lobe are generally quite tenuous and it is most unlikely that the alleged traction force could produce symmetrical centrifugal dilatation. It is probable that in collapsed and fibrosed lobes due to peripheral bronchial obstructions the proximal dilations are produced by the pressure of the atmosphere acting in the lumen when the external support of aerated alveoli has been removed.

That bronchiectasis may sometimes be of congenital origin is confirmed by the discovery of widely dilated bronchi and bronchogenic cysts in the lungs of stillborn infants. Recorded instances of these are, however, very few. More common are the anomalous collections of pulmonary tissue containing dilated bronchi, which are sometimes found in the mediastinum, pleural cavities or beneath the diaphragm. Sometimes an area of pulmonary parenchyma loses its connection with the remainder of the bronchial tree and acquires all its blood supply from the systemic circulation. Such sequestered tissue is usually buried in a lower lobe and when infection has caused its dilated bronchi to rupture into the normal portion of bronchial tree it may not be possible bronchographically to distinguish the condition from acquired bronchiectasis. Kartagener (1933) supports the view that bronchiectasis is sometimes congenital with his observation that among those

with complete transposition of the viscera a quarter also have bronchiectasis. This extraordinary frequency has been confirmed by others and it has been noted that many have a developmental deficiency of cranial air sinuses as well.

Clinical Features

Many adult sufferers from bronchiectasis ascribe the commencement of their pulmonary symptoms to some time during childhood. If a study is made of the age when such symptoms began in a large group of children it will be found that a quarter have had chest trouble since infancy and no less than 60 per cent. since before the age of three years. With such an early age of onset it is natural to suspect some congenital basis for the disease, but it must be realized that examination of the past medical history of these children with bronchiectasis always shows that they have been unduly prone to pneumonia, measles, pertussis and bronchitis accompanied by bronchial spasm. Not only is the incidence of these diseases normally greater in the young child, but the younger the child when they occur the narrower the bronchial calibre and the more readily will the lumen become occluded. In this country few cases of bronchiectasis result from the inhalation of a foreign body and bronchial obstruction by the enlarged glands of primary tuberculosis accounts for only a small proportion.

On physical examination children with bronchiectasis are a little below the average weight for age, though most have a good skin colour and give the appearance of being well nourished. They show surprisingly little concern about their symptoms, but tend to avoid much exertion as this induces coughing. The general body posture indicates hypotonia rather than vitality and about a quarter of the children reveal a little deformity of the thorax such as a pigeon chest, depressed lower sternum, scoliosis, kyphosis or Harrison's sulcus. Though almost all have a cough most of the year, only three-quarters ever expectorate sputum and usually it is just swallowed. Children acquire the habit of shallow breathing and appear to use the diaphragm little, because any forceful respiration tends to move retained secretions on to sensitive bronchial mucosa and provokes a distressing bout of coughing. About half the cases show an impaired percussion note and reduced air entry over the area of bronchiectatic lung. A similar proportion show rhonchi in the chest, but bronchial breath sounds are only found in about a quarter. The most constant abnormality is the presence of crepitations over the diseased area and these can be heard in about 80 per cent. of all children. An estimate of the vital

capacity usually shows it to be rather less than the average for the weight of the child.

Some degree of clubbing of the fingers occurs in about 40 per cent. of children. Haemoptyses are rare and occur in only 5 per cent. Chronic nasal discharge is present in almost all children with bronchiectasis and is usually quite as troublesome to the child as his cough. A bout of pneumonia around the diseased bronchi is a not infrequent occurrence and some children seem particularly prone to such episodes. Empyema, secondary pyogenic abscess and amyloid disease are all rare in childhood. A complication sometimes seen is an haemorrhagic nephritis. Modern drug therapy and the institution of postural drainage has greatly altered the prognosis of bronchiectasis and the severity of its complications. Even without surgery few children fail to reach adult life, probably less than 5 per cent.

Though the prognosis has improved, it is impossible yet to assess the amount of shortening to the duration of life than the presence of this chronic disease might entail.

In bronchography the skill acquired by practice is more important than the method, but when working with children it is particularly important that the latter should involve as little discomfort as possible. Below the age of eight years a general anaesthetic is required to abolish the cough reflex and either ether or pentothal is used. The latter may be used in combination with Flaxedil and the two have the advantage of a short but shallow anaesthesia coupled with good muscular relaxation, so that the vocal cords are toneless and the cough reflex is abolished. As soon as the child is sufficiently unconscious he is raised to the sitting position and iodised poppyseed oil is slowly injected through a nasal or pharyngeal catheter so that it flows through the relaxed vocal cords into the trachea. Some operators prefer to insert an endotracheal tube and instil the oil through a second fine catheter or one built into the wall of the tube. As the radio-opaque oil is flowing into the bronchial tree the thorax is inclined 45° from the vertical towards the side it is desired to outline and bent forward and back for about a minute in each position. The catheter is then withdrawn and the trunk inverted over the couch for another minute before lying the child flat and taking the first X-ray. The whole bronchial system on the appropriate side should be evenly coated with contrast medium in a few minutes and the films exposed without delay. A postero-anterior view and a lateral are taken for the right side, but on the left a first oblique view is better than a lateral. The average quantity of opaque oil used in children is 1 ml. per year of age. In older children, after premedication and a local

anaesthetic has been injected, the oil can be instilled into the trachea through a special curved needle inserted into the cricothyroid membrane. It is necessary to take special care that the point of the needle is in the lumen of the trachea and to precede the oil by a local anaesthetic to abolish the cough reflex; the drug used is usually amethocaine and $1\frac{1}{2}$ minims of a 2 per cent. solution is injected per year of age. The crico-thyroid route results in a higher proportion of good bronchograms; it is soon over and no anaesthetist is required. The technique is not without its risks and should never be used unless the skill and important details have been learned under the personal supervision of an expert and the operator has the confidence of the child.

In children the left lung is a little more often bronchiectatic than the right and the disease is bilateral in 60 per cent. of cases. The left lower lobe is more often involved than any other and next in frequency come the lingula, right middle and right lower lobes. The right upper lobe is affected almost twice as often as the left. The disease often affects more than one lobe, but only in 8 per cent. is the entire lung involved. A curious finding in children is the extent to which the anterior pulmonary segments are involved. They are affected more frequently and severely than the posterior ones. A common combination is the anterior segment of an upper lobe, the middle or lingula bronchi and the anterior, medial and possibly lateral segments of a lower lobe. This odd segmental distribution is difficult to explain on the basis of bronchopneumonia being the cause of the bronchiectasis, because in that disease the posterior bronchi are usually affected more severely. Material aspirated into the chest from the pharynx also tends to flow more readily into posterior bronchi than anterior. There is probably some simple physiological basis for the phenomenon of which we are not yet aware.

It was mentioned earlier that in order to preserve the clinical identity of bronchiectasis as something apart from a mere physical sign, the disease must be defined as one in which the bronchi are permanently or chronically dilated. It is often difficult to decide from bronchograms done on a single occasion the duration and severity of any bronchial deformity present. In Plate I, which is a left oblique bronchogram, the lower lobe is collapsed and its bronchi are widely dilated. The lingual lobe is a little reduced in volume and, though the peripheral portions of the bronchi are normal, the proximal parts are much wider than normal. In the presence of such parenchymal collapse it is always better to delay making a diagnosis of bronchiectasis until a second bronchogram can be done after some months of active

treatment. Plate II is the lateral view of a bronchogram done on the same child five months later and it can be seen that the lung tissue has re-expanded and bronchi returned to a normal calibre. Despite the apparent severity of the bronchial dilatation in the earlier bronchogram, this did not, by definition, constitute bronchiectasis, for it was neither chronic nor permanent. Most radiologists reporting on the first bronchogram would only remark that there was bronchial dilatation in association with pulmonary collapse. Had the same appearance been present at the time of the second bronchogram it would have been reasonable to suspect bronchiectasis and chronic inflammatory change in these areas, because in the vast majority of cases pulmonary collapse in childhood resolves within two months. Plate III has been included to show an example of gross bronchiectasis and to illustrate the greater extent and severity in the anterior pulmonary segments which is so often a feature of the disease in childhood. Plate IV is an operation specimen of a shrunken bronchiectatic lobe. Widely dilated and inflamed bronchi surrounded by much white fibrous tissue can be seen and the intervening collapsed parenchyma is diffusely infiltrated with chronic inflammatory cells which show up white in this photograph.

Treatment

There can be no doubt that where infected secretions are retained in deformed bronchial cavities the main endeavour in treatment must be to empty these of their contents and keep them free from such accumulations, so that the bronchial inflammation can subside and repair occur. This is difficult to achieve, because by the time the patient is seen there is usually a chronic purulent sinusitis which continually reinfects the lung. Treatment must therefore be directed to the nose and chest simultaneously. The child is made to sleep over a tipping frame placed on his bed so that secretions can drain from the affected bronchi and nasal mucus will not be inhaled into the chest during sleep. In severe cases, the mother must be taught to place her child over the frame, or invert the thorax over the side of a bed, and clap the chest two or three times a day to help the expectoration of secretions which accumulate while the child is upright. Besides this postural drainage the child should be taught breathing exercises which encourage the use of the diaphragm, for as mentioned earlier bronchiectatic children learn the habit of shallow respiration. When well performed these breathing exercises provoke coughing, expectoration and the emptying of the bronchial cavities, but as soon as the retained secretions have been brought up there

PLATE 1.—Left oblique bronchogram.

PLATE 2.—Lateral view of same patient as Plate 1. Five months later.

PLATE 3.—Gross bronchiectasis, note the greater involvement of the anterior pulmonary segments.

PLATE 4.—Operation specimen of a shrunk bronchiectatic lobe.

is little further distress. Before the child is placed over the tipping frame or breathing exercises are commenced, a warm saline mixture of common salt and sodium bicarbonate may be given to soften secretions and aid their expectoration. A sputum pot should always be used, as this demonstrates to the child the efficacy of the treatment. Where the nose and accessory air sinuses are chronically infected the child is instructed how to blow his nose effectively and inhalations of Friars balsam or menthol are prescribed, as they tend to loosen the nasal exudate and encourage it to flow away more easily. In most children it is necessary to give frequent antral lavages before there is much improvement in the condition of the maxillary antra.

The building up of the body's vitality and resistance is an integral part of the treatment of children with bronchiectasis. A full diet with vitamins and tonic supplements should be given, the child is encouraged to indulge in vigorous games, with the exception of swimming, and prolonged residence should be arranged at a school situated where the climate is good.

Surgery

When it is judged that a particular area of lung has been so damaged that it can never recover its normal functions and may cause prolonged ill-health, it should be removed. In children the remaining pulmonary parenchyma appears to compensate well by hypertrophy, and possibly hyperplasia, without the thoracic deformity and emphysema which sometimes results in adults. As the disease is so often bilateral and may be distributed among several lobes, two operations of segmental removal are often required. In cases where the disease is too extensive to be entirely removed great alleviations of symptoms may follow the removal of the worst parts.

Children stand thoracic surgery very well and are running about the ward within a few days. Their subsequent progress is also good, for most recover the slight retardation in growth they previously showed. The vital capacity also improves and often reaches a figure comparable

with normal controls. Children who have had a pneumonectomy do not make such marked improvement and tend to remain below average in weight, height and vital capacity, though thoracic deformity may be slight. Owing to the prolonged bronchitis and upper respiratory infection associated with bronchiectasis a cough generally persists in lesser degree for some months or even a few years.

Conclusion

If the bronchial deformity is permanent or at least chronic in bronchiectasis, what is the fate of those who have not the disease to such an extent as to warrant surgery or are too advanced for it? Observation of children with bronchiectasis over the past ten years has shown that this bronchial deformity very rarely disappears or becomes more severe. Spread of the disease to previously normal bronchi has been seen in very few cases and in all of these it has been in some way secondary to surgery. As children grow into adolescence the severity of their symptoms abates and in milder cases these clear completely, though subject to recurrence with colds and influenza. No one can at present say how long this general abatement of symptoms is likely to last. Persistent granulations or small erosions in the imperfectly epithelialized walls of chronic bronchiectatic cavities can result in haemopstyses, and indeed dry bronchiectasis is one of the commoner causes of haemopstysis in the adult. The published figures on the prognosis of bronchiectasis are discouraging, but most relate to times without the advances of modern chemotherapy and antibiotics. It may well be that those children with mild bronchiectasis now have a normal expectation of longevity, but it is feared that the slowly progressive course of fibrosis and emphysema and the ever-present risk of complications will still result in a premature demise of those too extensive for surgery.

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