if they increased in frequency or duration) would persuade me to accept such treatment—provided that there was no operable stenosis or reversible factor such as hypertension. Their benefit probably lasts only two or three months and certainly if they have no clinically impressive effect they should gradually be discontinued after such a period.

The one part of my own medicine to which I would closely adhere is the strict supervision and control of high blood pressure. If I were so affected I would try hard to be what is popularly known as a "compliant customer." Symptomatic or not, with signs of left ventricular strain or hypertrophy or not, I would accept a strict regimen because the achievement of normal blood pressure is usually possible nowadays without serious side effects. This, above all other mentioned factors, can enormously improve the outlook, in direct proportion to the level of control achieved. One would try to acquire a passive frame of mind, accepting the necessity of regular "check-ups": retinoscopy, ECG, and a chat with a trusted colleague without becoming too preoccupied or introspective. In fact, I would try to emulate the best of my own patients.

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Today's Treatment

Diseases of the respiratory system

Bronchiectasis and cystic fibrosis

MARGARET E HODSON

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Bronchiectasis

Bronchiectasis is characterised by dilatation of the bronchi. This dilatation may be generalised or confined to a single segment or lobe of the lung, and it may be tubular, fusiform (spindle shaped), or saccular (cystic).

Aetiology

Bronchiectasis usually starts in childhood but may begin in adult life. The commonest cause is damage to the bronchial tree after infections such as tuberculosis, bacterial or viral pneumonia, and bacterial infections complicating measles or pertussis. Bronchiectasis may be associated with certain congenital disorders—for instance, Kartagener's syndrome (a condition also characterised by dextrocardia and sinusitis), Williams-Campbell syndrome (bronchomalacia caused by congenital absence of cartilage distal to the first division of the peripheral bronchi), and sequestrated lung segments. Obstruction of the bronchi due to carcinoma, tuberculous stenosis, or inhaled foreign bodies may cause pulmonary collapse and subsequent infection leading to bronchiectasis. Recurrent sinus infections may also lead to it.

Bronchiectasis may complicate a more widespread disorder such as cystic fibrosis or some immunological deficiency such as hypogammaglobulinaemia. Bronchiectasis that characteristically affects the proximal bronchi may occur in allergic bronchopulmonary aspergillosis.

Brompton Hospital, London SW3 6HP
MARGARET E HODSON, MD, MRCP, senior medical registrar

Symptoms and diagnosis

The symptoms of bronchiectasis are cough and sputum production. There may be associated haemoptysis and the symptoms of a more generalised bronchitis such as wheeze and dyspnoea. The patient may have poor general health and finger clubbing. Crepitations may be heard over the areas of retained secretions. Bronchiectasis can often be diagnosed from the chest radiograph if a lateral and penetrated film are taken as well as the routine posterior-anterior view. Ring or tubular shadows may be seen, or abnormalities of the vascular pattern. The diagnosis can be confirmed and the full extent of the disease defined by bronchography, though this is an unpleasant investigation and is justified only if the patient is a candidate for surgical treatment.

Treatment

Treatment may be medical or surgical. If a patient has moderate or severe symptoms, is otherwise fit, has good lung function, and has bronchographic evidence of localised disease then he should be offered surgical resection of the affected lobe. If the remaining lung function is reasonable patients can stand resection of one lobe, the left lower lobe and lingula, or the right lower and middle lobes. In carefully selected cases excellent results can be obtained and a lifetime of medical treatment avoided. All these patients should receive intensive medical treatment before operation.

Most cases of bronchiectasis are unsuitable for surgery either because the patient has poor lung function or because the disease is widespread. The most important aspect of medical treatment is regular postural drainage of the affected segments or lobes. To drain the basal bronchi the patient may raise the foot of the bed; special tipping beds are available for this purpose. At home a patient may use a pile of pillows.

The middle lobe or lingula is drained with the patient on his back; the foot of the bed is raised, and a pillow placed under the affected side. The patient should maintain the appropriate posture for 10-15 minutes night and morning and during that time take deep breaths and cough to dislodge the secretions. If possible, a relative or friend should be shown how to percuss the chest to aid expectoration. Postural drainage is time consuming and embarrassing for the patient, and he often needs encouragement to continue with this routine. During acute exacerbations physiotherapy should be increased to four times daily.

The most important pathogens in the sputum of patients with bronchiectasis are *Haemophilus influenzae*, *Staphylococcus aureus*, and *Streptococcus pneumoniae*. Anaerobic organisms may also be important. All patients with bronchiectasis should have regular sputum samples cultured for aerobes and anaerobes. If a pathogenic bacterium is found then appropriate antibiotics should be given. Many *H influenzae* organisms are resistant to co-trimoxazole, so amoxycillin 500 mg four times a day for 10 days is the drug of choice in this infection; an alternative is tetracycline, 500 mg four times a day. If *Staph aureus* is isolated from the sputum cloxacillin, 500 mg four times a day, should be given. Anaerobic pathogens may be treated with metronidazole, 800 mg three times a day. Generally antibiotics should be given intermittently as their continuous use suppresses the normal respiratory tract flora and troublesome organisms such as *Pseudomonas aeruginosa* may emerge. Patients with bronchiectasis should have a supply of antibiotics at home so that they can take a 10-day course as soon as they develop an upper respiratory tract infection or their sputum increases in volume or becomes more purulent. Amoxycillin is suitable for this purpose.

Patients with large volumes of purulent sputum in whom simpler measures have failed should be admitted to hospital for intensive treatment. They should have intensive (four to six times daily) physiotherapy and either benzylpenicillin 600 mg four times a day and streptomycin 0·5 g twice a day intramuscularly for 10-14 days, or chloramphenicol 500 mg four times a day for 10 days. A few patients infected with *Ps aeruginosa* will benefit from a 10-day course of intravenous gentamicin, about 2 mg/kg three times a day (dose depending on blood levels), and carbenicillin 5 g four times a day. The sputum volume may decrease and the patient's general state of health improve, but the organism is rarely eliminated from the sputum.

A few patients with severe disease may benefit from long-term chemotherapy. Tetracycline, either 250 mg or 500 mg four times a day, for two days a week may be suitable. This should be continued only if the patient's condition definitely improves.

Patients who have allergic bronchopulmonary aspergillosis may need steroids to prevent further bronchial wall damage, and those with hypogammaglobulinaemia may benefit from gammaglobulin. Cystic fibrosis presents special problems, which will be discussed later.

All patients with bronchiectasis should be told not to smoke. Sepsis in the paranasal sinuses and teeth should be eliminated. All should have their forced expiratory volume in one second (FEV_1) and forced vital capacity (FVC) measured before and after taking salbutamol to see if they have any reversible airways obstruction. If they show improvement they should inhale salbutamol before performing their postural drainage. They should be offered immunisation against influenza each autumn, and they must have adequate standards of general nutrition and housing.

Complications

Complications include recurrent acute exacerbations of symptoms and pneumonia. Sinusitis is common and must be treated adequately. Haemoptysis is common and may be frightening. It is rarely severe, however, and usually clears with antibiotics for the underlying infection; if life-threatening it may call for surgery. Empyema, brain abscess, and amyloid are rare complications of bronchiectasis. Many patients progress to cor pulmonale after years of pulmonary sepsis and arterial hypoxaemia.

Prevention

All episodes of pulmonary infection and collapse must be treated adequately, especially in childhood. Children should be immunised against diseases such as pertussis. Anyone suspected of inhaling a foreign body must undergo bronchoscopy. Patients with allergic bronchopulmonary aspergillosis must be correctly diagnosed and treated at an early stage before bronchial wall damage becomes permanent.

Cystic fibrosis

Cystic fibrosis affects many exocrine glands throughout the body. The main clinical features are pancreatic insufficiency causing malabsorption (in 95% of patients); excess mucus production in the respiratory tract causing recurrent or chronic bronchopulmonary infection, which is the commonest cause of death in these patients; and a high sodium concentration in the sweat, providing a useful diagnostic test for the condition.

Cystic fibrosis is inherited as a Mendelian recessive and affects 1 in 2000 live births in a Caucasian population. In 1938 80% of babies born with this condition died within one year of birth, but now with improved paediatric care, an increased understanding of the importance of postural drainage, and the advent of antibiotics many patients are surviving into adult life. Physicians should therefore be familiar with the presentation and treatment of this disease as there are now many patients who have "outgrown" the paediatricians.

Presentation and diagnosis

The disease is usually diagnosed in childhood. Ten per cent of patients present soon after birth with meconium ileus; this is usually treated surgically. The remainder present during childhood with recurrent chest infections, symptoms of malabsorption, failure to thrive, or a combination of these features.

Cystic fibrosis is diagnosed in children when the results of two sweat sodium estimations are above 70 mmol/l; in adults this test is less reliable.

Treatment

The patients are advised to take a low-fat diet and regular pancreatic supplements. Pancreatin (Pancrex) powders are available for babies and older children and adults take such preparations as Pancrex V Forte, 6-10 capsules with each meal. Alternatives are nutrizyme and cotazym. All patients should have regular supplements of the fat-soluble vitamins.

From the time of diagnosis the parents must be shown how to perform twice-daily postural drainage and percussion of the child's chest. They must understand that if he is to have any chance of survival any excess mucus must be regularly drained from his respiratory tract. This regular physiotherapy must continue throughout life and be increased during acute exacerbations.

All patients should have sputum regularly cultured for aerobes and anaerobes, and whenever pathogens are isolated they should be treated accordingly. In early childhood the most important pathogen is *Staph aureus*. Later *H influenzae* may become prominent and in older patients the major pathogen is *Ps aeruginosa*.

Many paediatricians keep children with cystic fibrosis on regular antistaphylococcal drugs, such as cloxacillin, for the first two years of life. Thereafter it is probably best to maintain them on intermittent antibiotics. Antibiotics should be given

during upper respiratory tract infections and increases in sputum volume or purulence. The antibiotic is chosen according to the bacteria present in the sputum but the most commonly used are amoxycillin, tetracycline, and cloxacillin, 500 mg four times a day in each case. Cephalosporins, erythromycin, co-trimoxazole, and occasionally chloramphenicol or metronidazole may be indicated. If a patient is infected with *Ps aeruginosa* and the chest radiograph or measurements of respiratory function indicate deterioration then he must be admitted to hospital and given a 10-day course of gentamicin, about 2 mg/kg three times a day (dose monitored by blood levels), and carbenicillin 5 g four times a day. This may greatly improve his condition but the organism is rarely eradicated from the sputum.

Many patients with cystic fibrosis have reversible airways obstruction. For this they should have salbutamol regularly at the time of postural drainage. Salbutamol (5 mg) 1 ml plus 2 ml of physiological saline is nebulised in either a Bird nebuliser or an Inspiron Mini-Neb nebuliser. These nebulisers can be driven by an oxygen supply or, more satisfactorily for home use, by an air compressor. The PGS Compressor with a Bird nebuliser is perhaps the best currently available combination. Positive-pressure ventilation should not be used in a patient with cystic fibrosis as there is an increased risk of pneumothorax. In a few patients with gross airways obstruction steroids may be indicated but these should only be continued if definite benefit is obtained.

There is now no place for mist tents or mucolytics in treating cystic fibrosis, but during acute exacerbations some patients find it easier to expectorate if an ultrasonic nebuliser provides humidification.

These patients must not smoke and should be offered influenza vaccinations each autumn. All patients should be under regular outpatient supervision. At the outpatient clinic they should receive treatment from a professional physiotherapist; sputum should be taken for culture, and peak expiratory flow rate, FVC, and FEV_1 should be measured. Chest radiographs should be taken at least every six months, and the patient should be weighed at each visit. Weight loss should alert the clinician to a deterioration of lung function, development of diabetes mellitus, or increasing malabsorption.

Complications and their treatment

Meconium ileus and "meconium ileus equivalent"—Meconium ileus presents as intestinal obstruction soon after birth. The baby must be rehydrated and usually surgical treatment is indicated. "Meconium ileus equivalent" is intestinal obstruction occurring in the adult when tenacious mucus and faecal matter obstruct the small bowel. The treatment of choice is medical (with acetylcysteine[1]), but occasionally surgery is required.

Deterioration of lung function—If there is evidence of deterioration in lung function, either an acute exacerbation of symptoms or a decrease in respiratory function values, the patient should be admitted for intensive physiotherapy and a course of whatever antibiotics are indicated by the results of sputum culture.

Haemoptysis is a common problem. Usually it is transient and reassurance is all that is needed. It may be caused by increased infection and therefore antibiotic treatment should be reviewed. Occasionally it is life-threatening, and the bronchial arteries have to be ligated.

Pneumothorax occurs more often in adults than in children, and it may be bilateral. Any patient with cystic fibrosis who complains of chest pain must have a chest radiograph to exclude this complication. A very small pneumothorax may require no treatment, but most do need treatment as lung function is already reduced. An intercostal tube should be inserted and connected to an underwater seal. Suction may be applied but often the lung does not re-expand or the pleura may fail to become adherent. If the patient is fit enough he should have pleurectomy; in a very debilitated patient chemical pleurodesis may be attempted.

Diabetes mellitus occurs in some of the older patients. It is often controlled by diet and oral hypoglycaemic agents but may require insulin.

Rectal prolapse is a problem in the toddler. Under sedation a lubricated gloved finger can usually replace the prolapsed rectum. Surgery is rarely necessary.

Nasal polyps and sinusitis—Polyps that are symptomatic should be removed and sinusitis should be treated by antibiotics and nasal decongestants, and antral lavage if indicated.

Liver disease—Only a small proportion of older patients develop clinically important features of portal hypertension due to cirrhosis. They may present with bleeding varices, hypersplenism, or hepatosplenomegaly. Treatment should be conservative whenever possible. Shunt operations have been performed but the mortality is considerable. Vitamin K_1 is indicated if the prothrombin time is prolonged.

Reproduction—Most men are infertile owing to abnormalities of the vasa deferentia. Women are fertile, but during pregnancy lung function often deteriorates as postural drainage is more difficult to perform in late pregnancy. Women with cystic fibrosis contemplating pregnancy should discuss this very carefully with their medical advisers.

Cardiac failure—The progressive pulmonary disease inevitably leads to cor pulmonale and overt cardiac failure. This should be treated with diuretics, potassium supplements, and digoxin as indicated.

Management of associated social problems

The paediatrician who diagnoses the condition is responsible for giving adequate genetic counselling to the parents. In each future pregnancy there is a one in four chance of another affected child, a one in two chance of a carrier, and only a one in four chance of a baby who is neither affected nor a carrier.

Most children with cystic fibrosis can attend normal schools; because of their physical disability their future careers should be planned after discussion with the doctor and hospital social worker.

Patients who plan to get married should be encouraged to bring their future partner to the clinic so that the various problems associated with cystic fibrosis can be discussed.

Death

Despite the best available treatment most patients die before they are 40. Many die during their teens and 20s. When a patient with cystic fibrosis has evidence of steadily deteriorating lung function, despite intensive treatment, there is no justification for mechanical ventilation. He should be treated symptomatically. The best management of a young adult suffering from a terminal illness requires the combined skills of the doctor, nurses, physiotherapists, social workers, and the hospital chaplain.

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