EPIDEMIC OBSERVATION UNIT

Cystic fibrosis in childhood

The disease, which is genetically determined, is probably due to the secretion of an abnormally viscid mucus in the lungs and gastro-intestinal tract. The incidence of the disease is thought to be of the order of one in every 2,000 live births in Britain, but it is not accurately known.

About one child in ten with cystic fibrosis presents with meconium ileus resulting in intestinal obstruction in the neonatal period. The remainder present with respiratory and gastro-intestinal symptoms at any time in childhood.

In respect of the respiratory system there is usually a history of recurrent chest infection; the individual attacks are often unusually prolonged and slow in response to therapy. Some clubbing of the fingers may develop early, and radiographic changes in the chest also usually develop early with evidence of patchy consolidation and collapse. Physical signs in the chest occur, but usually rather later in the disease. With the progression of respiratory disease the chest generally becomes barrel shaped with bowing of the sternum.

The earliest gastro-intestinal sign is usually poor weight gain in spite of an excellent appetite. The most characteristic sign is the frequent passage of stools, which tend to be large, light in colour, greasy and offensive. Rectal prolapse is not infrequent. Abdominal pain and vomiting do not usually occur. On examination the abdomen is often somewhat distended and tympanitic. In the later stages the liver is almost always palpable; very occasionally older children may present with portal hypertension.

If it is suspected on clinical grounds that a child has cystic fibrosis he or she is best referred to a paediatrician for laboratory confirmation of the diagnosis. There is a low level of tryptic activity in duodenal juice, and a raised level of sodium and chloride in sweat. The earlier the diagnosis is made the greater the opportunity to prevent permanent lung damage.

Where there has been difficulty in establishing the clinical diagnosis it is usually because *only* the gastro-intestinal or *only* the respiratory system appeared to be affected. In the former instance, the primary diagnosis was often that of coeliac disease or fat intolerance, and the latter instance, that of bronchitis, bronchiectasis or asthma.

The condition is genetically recessive, so that it is not uncommon to find brothers and sisters affected; once parents have had one affected child the risk of recurrence in later children is one in four.

CORRECTION

In Dr Illingworth's article *The Choice of Occult Blood Tests in General Practice*, the sentence on page 36, line 11, of *Journal* 48 should read: " It is likely that more will be heard of this technique, and not only because of its safety."