key points

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Cystic fibrosis (CF) is a multisystem genetic disorder affecting approximately 1 in 2,500 live births in the UK. Although all newborns undergo screening for CF, around 15% of infants will present shortly after birth with meconium ileus and some will already have faltering growth when the screening results are available at 3-4 weeks of age. Infants who present with a meconium ileus should be treated with a high index of suspicion for CF until proven otherwise. Around 93% of children are diagnosed in the first three months of life and another 5% before the age of 12 months.

Mucociliary dysfunction leads to accumulation of mucus in the airways and secondary infection. Respiratory symptoms may be non-specific initially and include cough and wheeziness, frequent respiratory infections and, in older children, sinusitis. Pancreatic function is impaired in most children, and symptoms of malabsorption include meconium ileus, steatorrhoea, faltering growth, distended abdomen and constipation.

Distal intestinal obstruction syndrome is a serious complication that presents with acute abdominal pain and constipation. Liver disease may result from fatty replacement or interstitial fibrosis due to inspissated bile. CF related diabetes becomes prevalent in the teenage years, often presenting with poor weight gain in a young person who had previously been thriving.

Most newly diagnosed infants with CF will commence once daily physiotherapy for airway clearance. At first this is delivered by their parents but as children get older they will increasingly be in charge of their airway clearance. Young children are vulnerable to respiratory infection with *Staphylococcus aureus* and, in the UK, most receive a prophylactic antibiotic, commonly flucloxacillin, until the age of three years. Exacerbations (worsening cough which is wet or associated with sputum) must be treated proactively with antibiotics and increased airway clearance. Children with CF should have antibiotics when a wet cough has been present for more than a week even if there is little to hear on auscultation of the chest. The CF centre should be contacted for advice on the choice of oral antibiotics and whether intravenous antibiotics might be needed. Pancreatic enzyme replacement therapy has been shown to improve fat absorption with concomitant improvement in calorie intake. Supplements of fat-soluble vitamins (A, D, E and K) may also be necessary as these vitamins are poorly absorbed.