

Cystic fibrosis

europeanlung.org/cystic-fibrosis/

Cystic fibrosis (CF) is an inherited disease, which is most common in white children and young adults, although it can affect people of any race. It used to be thought of as a disease of the lungs and digestive system, but it is now known to affect most organs in the body.

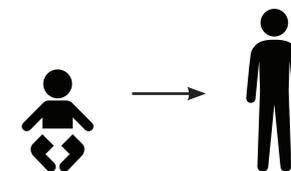


Areas for action

- There is a need to ensure that specialised services for adults with CF are established in all European countries and that they offer standards of care similar to paediatric clinics
- A dedicated CF unit is essential for best care
- CF should be diagnosed early, preferably by newborn screening
- As there will be an increased demand for lung transplantation, organ donation should be promoted to ensure that supply can meet the demand
- Treatment has in the past been solely aimed at symptoms of CF. Future research needs to concentrate on correcting the underlying abnormalities - the first treatment that corrects the basic defect has been developed (Kalydeco™ (Ivacaftor, VX-770)) for use in CF due to one specific gene variant
- Patients should be genotyped to allow for better targeted treatments



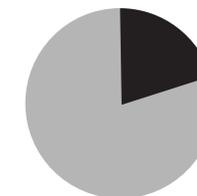
CF is an **inherited (genetic) condition**; a large number of variants of the faulty gene have been identified and the severity of the condition is dependent on the variation



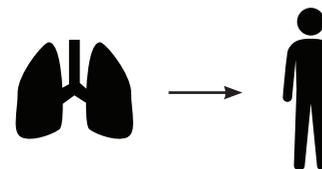
CF is **changing from a disease of childhood into a disease of adults**. Today, 42% of CF patients are aged >18, 5% >40% years



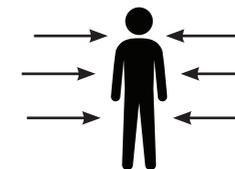
Newborn screening helps reduce prevalence, because parents are then able to make better informed decisions



15% of patients are **hospitalised** at least once a year



0.6% of CF patients have an **organ transplant** each year but this number is increasing; in most transplant centres CF is now the commonest reason for lung transplantation



Although CF has a major impact on the lungs **several other systems of the body are also affected** and non-respiratory complications are becoming more prominent as life expectancy increases