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NHS urged to find money for cystic fibrosis drug

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The head of the NHS has been urged to approve a drug that could add years to the lives of people with cystic fibrosis, an inherited disease that typically kills patients before they reach middle age.

The disorder, which affects 10,800 people in Britain, is caused by genetic mutations that disrupt their cells' ability to allow water and salt in and out. This leads to a build-up of mucus in their organs that can trap deadly infections and compromise their breathing.

About half of the cystic fibrosis patients in the UK have a mutation that means their lives could be extended through a drug called Orkambi, one of the first therapies to have a serious effect on its underlying causes.

The drug was rejected last year by the government's medicines evaluation agency because of its price, £104,000 a year for each patient, and a lack of information about its long-term benefits. New data, however, shows that Orkambi may halt or reverse lung damage in children aged under 12. It has also been found to slow the loss of lung function in older patients by 42 per cent.

The Cystic Fibrosis Trust is lobbying the health service to come back to the negotiating table with Vertex, the drug's manufacturer, after a ten-year deal in Ireland that is thought to have resulted in a substantial discount.