

Data request 269

Supporting information for NHS Health Technology Appraisals

Data taken from the UK Cystic Fibrosis Registry 2015 dataset. To protect the identity of individuals, numbers of less than 5 have been suppressed.

Gating mutations

Number of individuals with at least one copy of (1) G551D and (2) another gating mutation covered by the European marketing authorisation for ivacaftor use in age ranges: <2; 2-5; and ≥6 in each nation of the UK, by centre attended.

	England			Scotland			NI			Wales		
	<2	2-5	≥6	<2	2-5	≥6	<2	2-5	≥6	<2	2-5	≥6
G178R	0	<5	16	0	0	0	0	0	0	0	0	0
S549N	0	<5	23	0	0	0	0	<5	0	0	0	<5
S549R	0	0	8	0	0	<5	0	0	0	0	0	<5
G551D	16	43	350	<5	6	76	<5	<5	30	0	<5	12
G551S	0	0	<5	0	0	0	0	0	0	0	0	0
G1244E	0	0	0	0	0	0	0	0	0	0	0	0
S1251N	0	0	0	0	0	0	0	0	0	0	0	0
S1255P	0	0	0	0	0	0	0	0	0	0	0	0
G1349D	0	0	0	0	0	0	0	0	0	0	0	0

R117H

Number of individuals with at least one copy of a R117H mutation in age ranges: <18; and ≥18 in each nation of the UK, by centre attended.

England (n=310)		Scotland (n=56)		NI (N=54)		Wales (N=13)	
<18	≥18	<18	≥18	<18	≥18	<18	≥18
147	163	24	32	23	31	5	8

Homozygous F508del

Number of individuals with two copies of the F508del mutation in age ranges: <6; 6-11; and ≥12 in each nation of the UK, by centre attended.

England			Scotland			NI			Wales		
<6	6-11	≥12	<6	6-11	≥12	<6	6-11	≥12	<6	6-11	≥12
588	639	2831	39	67	243	23	28	101	20	31	118