## Cystic Fibrosis a fight we must win

## Number of individuals eligible by genotype for CFTR modulating therapy in each nation of the UK, defined by centre attended

These data are taken from the UK CF Registry verified data set for 2018 containing 9525 people in the UK with annual reviews. All tables exclude those who have ever had a lung transplant. Totals below five people are suppressed.

### 1. Individuals with one or two of the mutations G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N or S549R

	Age at 2018 annual review								
UK nation by	<2	<2 ≥2 and <6 ≥6 and <12 ≥12 and ≥18 years All ages							
centre	years	years	years	<18 years					
England	15	61	65	62	286	489			
Northern Ireland	<5	<5	7	5	23	41			
Scotland	<5	<5	13	14	48	81			
Wales	0	<5	<5	<5	16	21			
Total	22	68	-	-	373	632			

#### 2. Individuals with one or two of R117H mutation

	Age at 2018 annual review						
UK nation by centre	<2 years	≥2 and <6 years	≥6 and <12 years	≥12 and <18 years	≥18 years	All ages	
England	21	54	89	40	198	402	
Northern Ireland	0	8	8	5	35	56	
Scotland	0	8	14	8	35	65	
Wales	0	0	0	<5	11	-	
Total	21	70	111	-	279	-	

#### 3. Individuals who are homozygous for the F508del mutation

	Age at 2018 annual review							
UK nation by	<2	<2 ≥2 and <6 ≥6 and <12 ≥12 and ≥18 years All a						
centre	years	years	years	<18 years				
England	124	449	703	625	2100	4001		
Northern Ireland	10	17	35	22	70	154		
Scotland	8	32	50	50	191	331		
Wales	6	15	40	25	89	175		
Total	148	513	828	722	2450	4661		

# 4. Individuals with one copy of DF508del and one of the following CFTR mutations P67L, R117C, L206W, R352Q, A455E, D579G, 711+3A $\rightarrow$ G, S945L, S977F, R1070W, D1152H, 2789+5G $\rightarrow$ A, 3272 26A $\rightarrow$ G, and 3849+10kbC $\rightarrow$ T

	Age at 2018 annual review							
<b>UK nation by</b>	<2	≥2 and <6	≥6 and <12	≥12 and	≥18 years	All ages		
centre	years	years	years	<18 years				
England	18	37	51	30	250	386		
Northern Ireland	<5	6	<5	<5	21	33		
Scotland	<5	<5	<5	<5	44	55		
Wales	0	0	7	<5	5	-		
Total	21	-	66	37	320	-		

## 5. Individuals with one copy of DF508del and one of the CFTR mutations eligible for 'triple therapies' VX445 or VX659

	Age at 2018 annual review					
UK nation by centre	<2 years	≥2 and <6 vears	≥6 and <12 vears	≥12 and <18 years	≥18 years	All ages
England	36	140	210	193	698	1277
Northern Ireland	<5	7	15	12	26	-
Scotland	<5	16	19	20	76	-
Wales	<5	11	13	16	48	-
Total	-	174	257	241	848	-

#### 5a. Individuals with at least one copy of DF508del and therefore eligible for Ivacaftor-Tezacaftor-Elexacaftor

	Age at 2018 annual review							
UK nation by	<2 ≥2 and <6 ≥6 and <12 ≥12 and ≥18 years All a							
centre	years	years	years	<18 years				
England	232	825	1233	989	3849	7128		
Northern Ireland	15	43	68	45	187	358		
Scotland	18	65	105	101	419	708		
Wales	8	31	65	48	195	347		
Total	273	964	1471	1183	4650	8541		