



IP:

lgG

input

IP:

dyskerin TCAB1

IP:





	DAPI	IF:53BP1	FISH:Telomere	Merge
siCTRL				
siTIN2		L. S. C. S. S.		4 Jane
siTPP1				
siPOT1				







В

А



siCtrl

siTIN2

Nuclei with >5 TIFs (n)								
siCtrl				siTIN2				
GFP	5 (144), 3.5%	n.s.		GFP	46 (127), 36.2%	n.s.		
OB-WT	6(126), 4.8%	n.s.		OB-WT	41 (111), 36.9%	n.s.		
OB-RR	4 (113), 3.5%	n.s.		OB-RR	39 (116), 33.6%	n.s.		

Е

D



В





В

		% of
	Associated	Catalytic
Mutation in TERT	disease	function
P33S	IPF	~100%
L55Q	IPF	~40%
G100V	-	25%
V144M	IPF	~80%-100%
A202S	AA	<20%
R979W	AA/DC	<10%
V1090M	AA	<20%
V1110M	IPF	~50%
E1116fsX	IPF/AA	~10%
F1127L	HH/DC	-
1127NAA(C-DAT)	-	~100%