

Supplementary Information

Generation of novel patient-derived *CIC-DUX4* sarcoma xenografts and cell lines

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Table S1. STR analysis

Microsatellite (Chromosome)	NCC-CDS1-X1- C1 (P14)	NCC-CDS1-X2- C1 (P35)	NCC-CDS1- X1 (PDX)	NCC-CDS1- X2 (PDX)	Tumor Tissue
Amelogenin (X Y)	X,X	X,X	X,X	X,X	X,X
TH01 (3)	7,9	7,9	7,9	7,9	7,9
D21S11 (21)	30,30	30,30	30,30	30,30	30,30
D5S818 (5)	10,12	10,12	10,12	10,12	10,12
D13S317 (13)	11,12	11,12	11,12	11,12	11,12
D7S820 (7)	12,13	12,13	12,13	12,13	12,13
D16S539 (16)	9,13	9,13	9,13	9,13	9,13
CSF1PO (5)	11,13	11,13	11,13	11,13	11,13
vWA (12)	14,16	14,16	14,16	14,16	14,16
TPOX (2)	8,8	8,8	5,8	8,8	8,8

Cells were compared to a normal and tumor tissue from the patient.

P indicates a passage number of a cell line.

Table S2. KEGG pathways enriched in the proteome of different samples.

KEGG Enrichment Rank	Pathway	Primary tumour	NCC-CDS1-X1	NCC-CDS1-X3	X1-C1	X3-C1	Observations
1	Proteasome	1	2	2	3	3	Consistently enriched in all samples
2	Spliceosome	2	3	3	2	2	
3	Ribosome	3	1	1	1	1	
4	Pathogenic Escherichia coli infection	4	4	5	5	7	
5	Glycolysis / Gluconeogenesis	5	6	4	8	8	
8	Pyruvate metabolism	8	8	9	9	9	
9	Huntington's disease	9	15	13	6	6	
12	Pentose phosphate pathway	12	7	6	15	21	
14	Citrate cycle (TCA cycle)	14	5	7	4	4	
19	Glyoxylate and dicarboxylate metabolism	19	9	11	22	12	
17	Aminoacyl-tRNA biosynthesis	17		16	7	5	
18	Parkinson's disease	18		15	10	10	
24	Alzheimer's disease	24		20	11	13	
6	Focal adhesion	6	10	21			Enriched in all but xenograft tissues and cell lines
25	Regulation of actin cytoskeleton	25	12				
23	Tight junction	23		14			
15	Fatty acid elongation in mitochondria	15			16	16	Enriched in xenograft tissues and cell lines
16	Fatty acid metabolism	16			20	11	
20	Methane metabolism	20				20	
29	Propanoate metabolism		11		17		Irregularly enriched in primary tumours and cell lines
35	DNA replication			17	18		
33	Cysteine and methionine metabolism			8		19	
7	Complement and coagulation cascades	7					Enriched only in primary tumor tissue
10	Systemic lupus erythematosus	10					
11	Prion diseases	11					
13	Antigen processing and presentation	13					
21	Valine, leucine and isoleucine degradation	21					
22	Viral myocarditis	22					
27	Gap junction		14	12			Enriched only in xenograft tissues
30	Fructose and mannose metabolism		13	10			
31	Cell cycle		16	18			
32	Leukocyte transendothelial migration		17	25			
36	One carbon pool by folate			19			
45	Lysine degradation				13	14	Enriched only in cell lines, either consistently or irregularly
46	Valine, leucine and isoleucine biosynthesis				14	24	
48	Arginine and proline metabolism				21	22	
28	Fructose and mannose metabolism				19		
41	Oocyte meiosis				23		
44	Oxidative phosphorylation				12		
47	Butanoate metabolism				18		
38	Cardiac muscle contraction					23	
39	Protein export					17	
51	Oxidative phosphorylation					15	
52	Selenoamino acid metabolism					25	

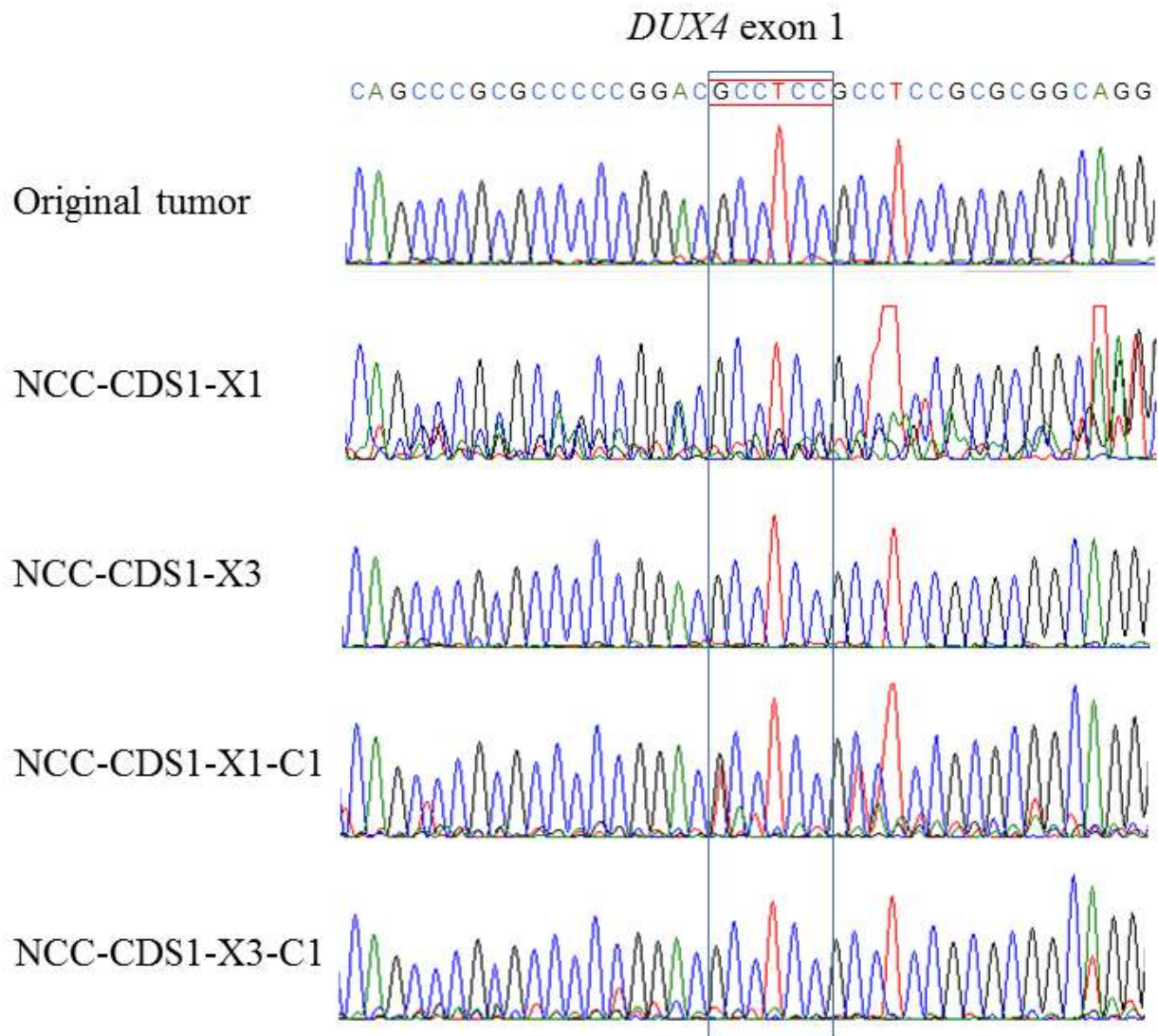


Figure S1. Sanger sequencing showed that the nucleotide sequence 5'-CGCCTC-3' was detected within *DUX4* exon 1, but not *DUX4L*, indicating that *CIC* was fused to *DUX4* (4q35). The sample names are shown on the left side of the panels.

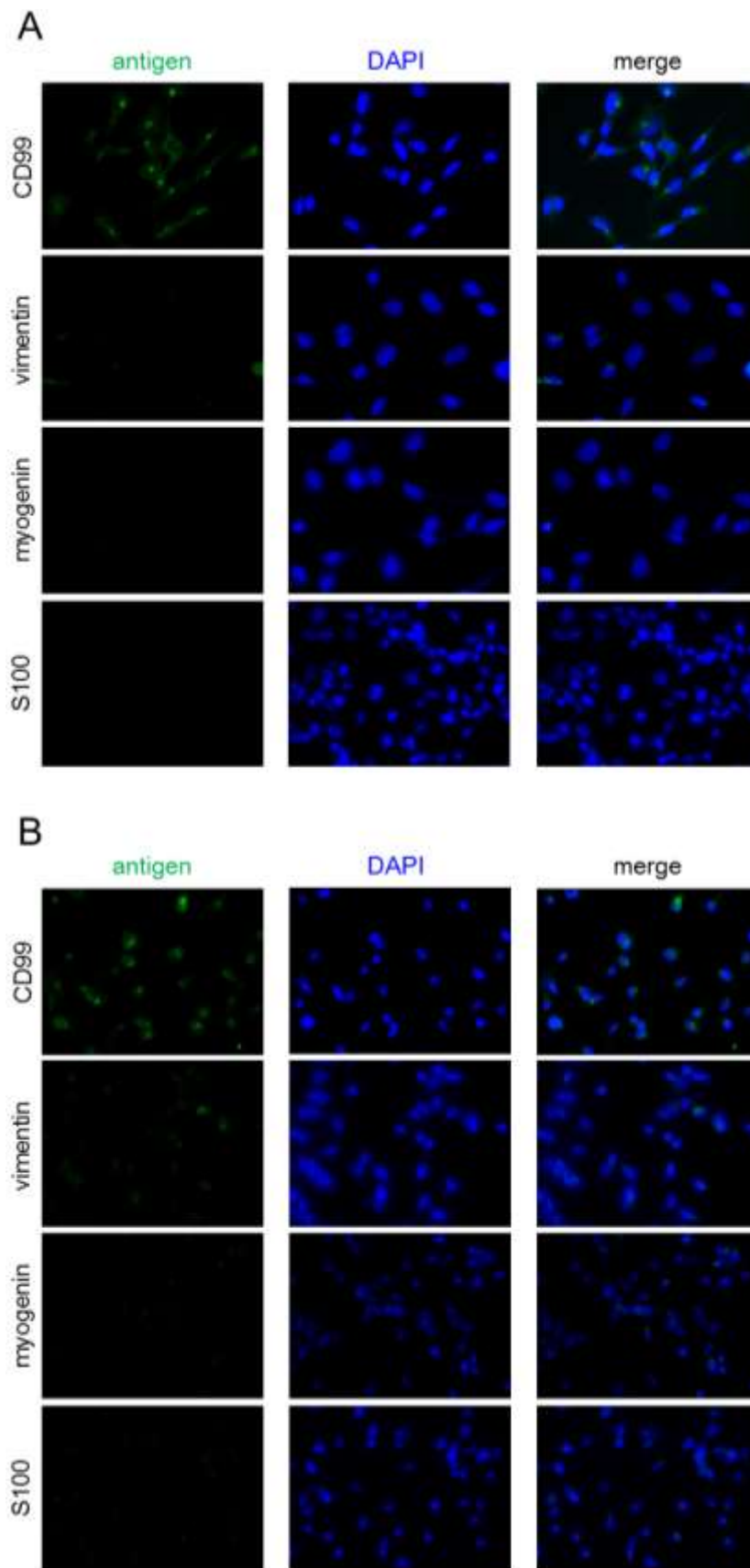


Figure S2. Immunofluorescence study for CD99, vimentin, myogenin, and S100. (A) NCC-CDS1-X1-C1, (B) NCC-CDS1-X3-C1.

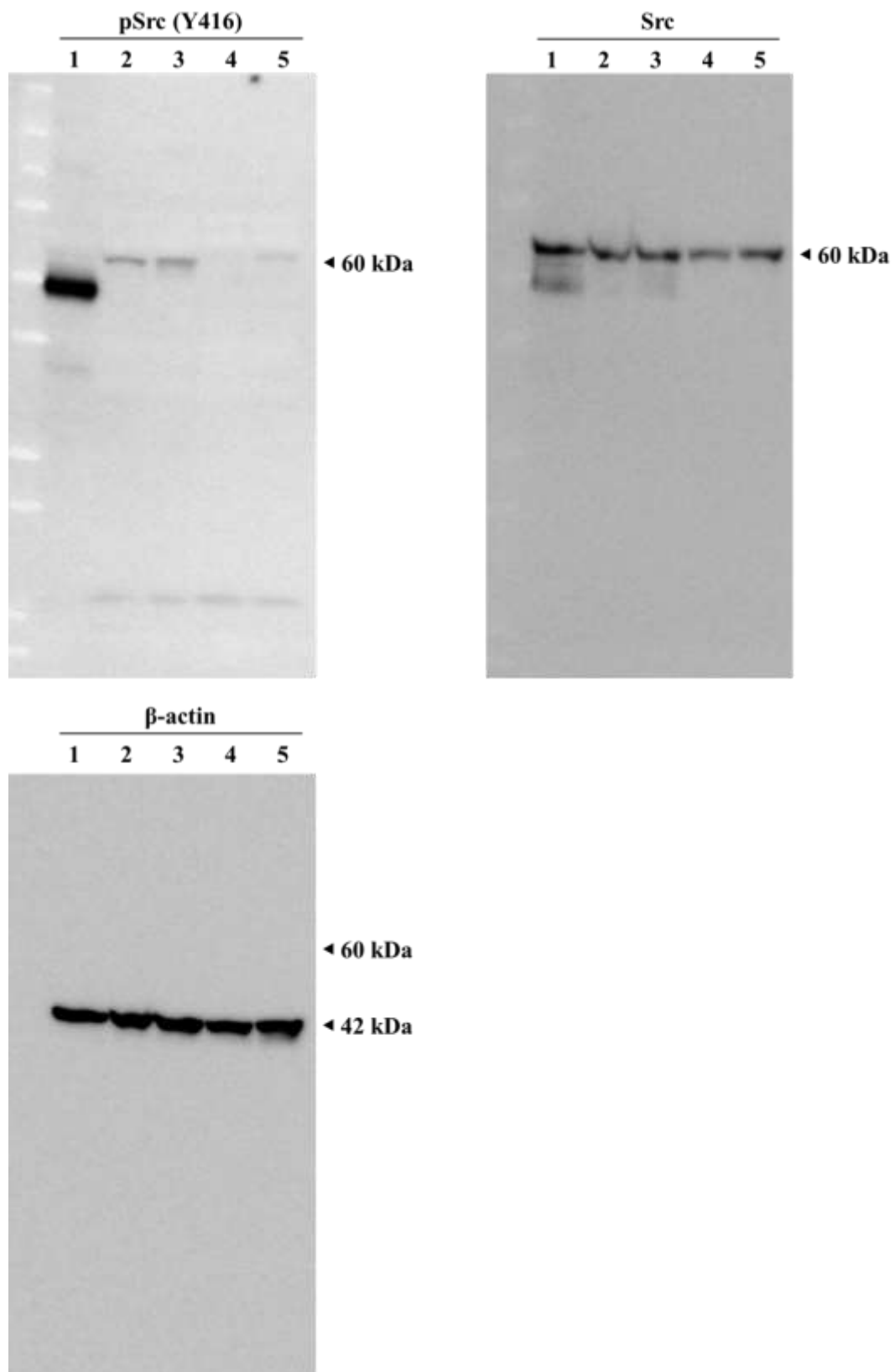


Figure S3. Phosphorylated Src expressions were analysed by Western blotting. Arrowheads indicate the position of Src with a molecular weight of 60 kDa. Lanes 1: tumour tissue, 2: NCC-CDS1-X1 tumour tissue, 3: NCC-CDS1-X3 tumour tissue, 4: NCC-CDS1-X1-C1 cells, 5: NCC-CDS1-X3-C1 cells.