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 | XI-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     |       |                                                    |
| 4                     | Pathogenic Escherichia coli infection                       | 4              | 4           | 5           | 5     | 7     |                                                    |
| 5                     | Glycolysis / Gluconeogenesis                                 | 5              | 6           | 4           | 8     | 8     |                                                    |
| 8                     | Pyrimidine metabolism                                        | 8              | 8           | 9           | 9     | 9     |                                                    |
| 9                     | Huntington's disease                                         | 9              | 15          | 13          | 6     | 6     |                                                    |
| 12                    | Fatty acid elongation in mitochondria                        | 12             | 7           | 6           | 15    | 21    | Consistently enriched in all but NCC-CDS1-X1 samples |
| 14                    | Fatty acid metabolism                                       | 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    |       | Consistently enriched in all but xenograft tissues and cell lines |
| 24                    | Alzheimer's disease                                          | 24             | 20          | 11          | 13    |       |                                                    |
| 6                     | Focal adhesion                                               | 6              | 10          | 21          |       |       | Enriched only in xenograft tissues                |
| 25                    | Regulation of actin cytoskeleton                             | 25             | 12          |             |       |       |                                                    |
| 23                    | Tight junction                                               | 23             |             |             | 14    |       | Enriched in xenograft tissues and cell lines       |
| 15                    | Fatty acid elongation in mitochondria                        | 15             | 16          | 16          |       |       |                                                    |
| 16                    | Fatty acid metabolism                                        | 16             |             | 20          | 11    |       |                                                    |
| 20                    | Methane metabolism                                          | 20             |             | 20          |       |       |                                                    |
| 29                    | Propanoate metabolism                                       | 29             | 11          | 17          |       |       |                                                    |
| 35                    | DNA replication                                             | 35             | 17          | 18          |       |       | Enriched only in primary tumour                   |
| 33                    | Cysteine and methionine metabolism                           | 33             | 8           |             | 19    |       |                                                    |
| 7                     | Complement and coagulation cascades                          | 7              |             |             | 7     |       |                                                    |
| 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                    | Clap junction                                               | 27             | 14          | 12          |       |       | Enriched only in xenograft tissues                |
| 30                    | Fructose and mannose metabolism                             | 30             | 13          | 10          |       |       |                                                    |
| 31                    | Cell cycle                                                  | 31             | 16          | 18          |       |       |                                                    |
| 32                    | Leukocyte transeendothelial migration                       | 32             | 17          | 25          |       |       |                                                    |
| 36                    | One carbon pool by folate                                  | 36             |             |             | 19    |       |                                                    |
| 45                    | Lysine degradation                                          | 45             | 13          |             | 14    |       |                                                    |
| 46                    | Valine, leucine and isoleucine biosynthesis                | 46             |             | 14          | 24    |       |                                                    |
| 48                    | Arginine and proline metabolism                             | 48             |             |             | 21    | 22    |                                                    |
| 28                    | Fructose and mannose metabolism                             | 28             |             |             | 19    |       |                                                    |
| 41                    | Oocyte meiosis                                              | 41             |             |             | 23    |       |                                                    |
| 44                    | Oxidative phosphorylation                                  | 44             |             |             | 12    |       |                                                    |
| 47                    | Butanoate metabolism                                       | 47             |             |             | 18    |       |                                                    |
| 38                    | Cardiac muscle contraction                                  | 38             |             |             |       | 23    |                                                    |
| 39                    | Protein export                                             | 39             |             |             |       | 17    |                                                    |
| 51                    | Oxidative phosphorylation                                  | 51             |             |             |       | 15    |                                                    |
| 52                    | Selenoamino acid metabolism                                | 52             |             |             |       | 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 SuppleTable S2, 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.