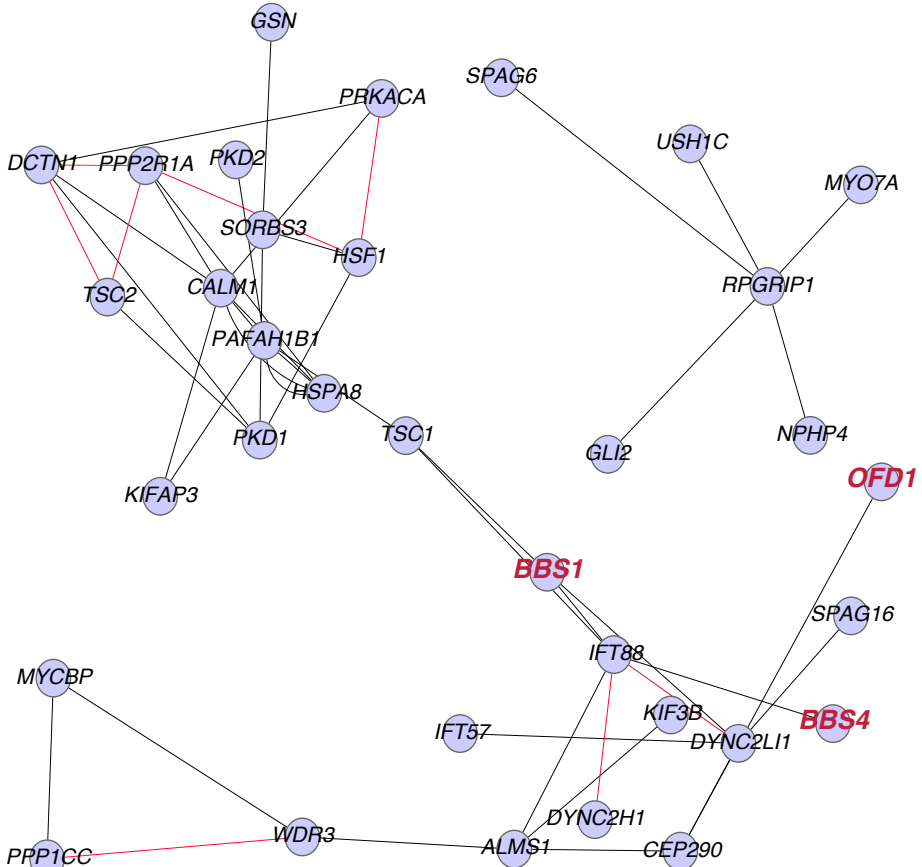
GSN
PRKACA
SPAG6
DCTN1
PPP2R1A
PKD2
USH1C
MYO7A
SORBS3
HSF1
RPGRIP1
TSC2
CALM1
PAFAH1B1
HSPA8
PKD1
TSC1
NPHP4
GLI2
KIFAP3
OFD1
BBS1
SPAG16
IFT88
MYCBP
KIF3B
BBS4
IFT57
DYNC2LI1
DYNC2H1
WDR3
ALMS1
CEP290
PPP1CC

Figure S1

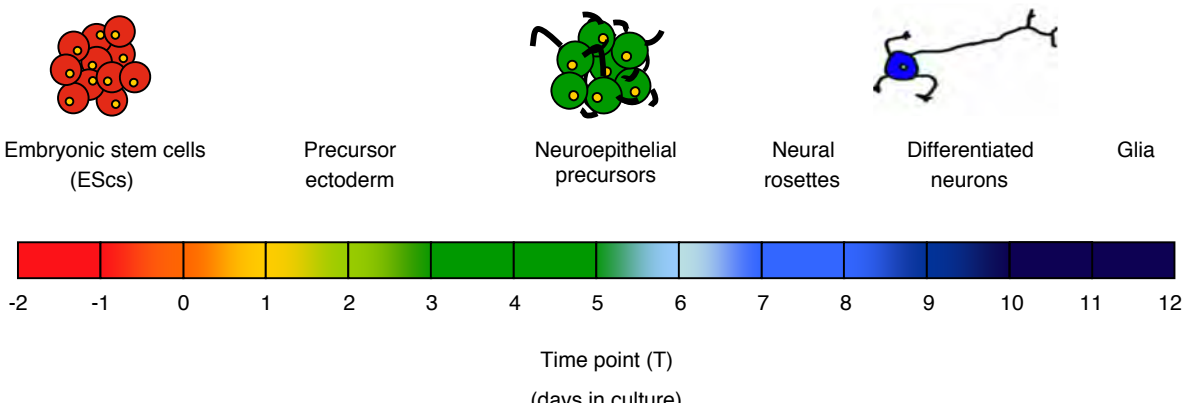
Embryonic stem cells
(EScs)
Precursor
ectoderm
Neuroepithelial
precursors
Neural
rosettes
Differentiated
neurons
Glia
-2
-1
0
1
2
3
4
5
6
7
8
9
10
11
12
Time point (T)
(days in culture)

Figure S2

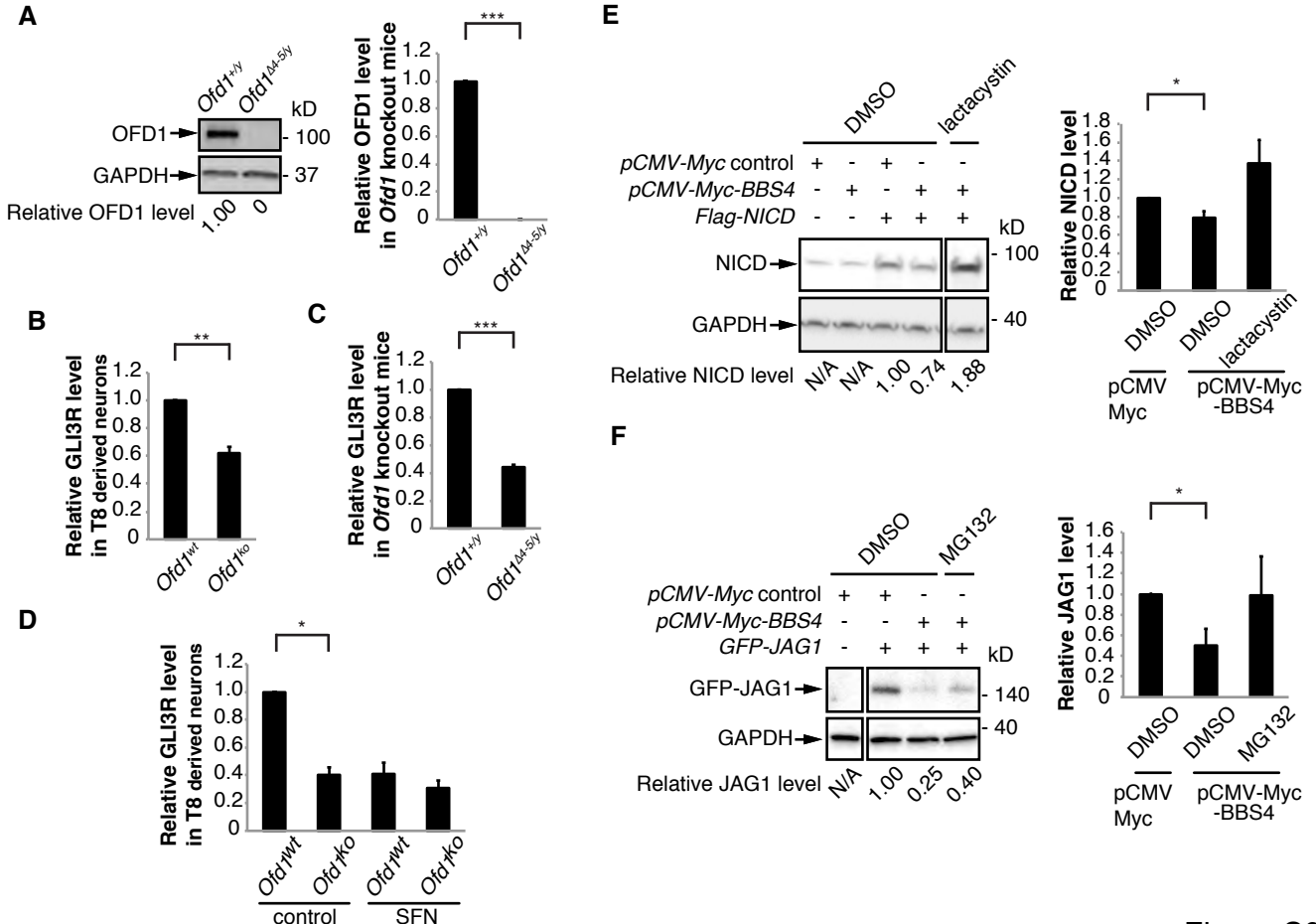
A
Ofd1+/y
Ofd1Δ4-5/y
kD
OFD1
100
GAPDH
37
Relative OFD1 level
1.00
0
Relative OFD1 level in Ofd1 knockout mice
***
B
Relative GLI3R level in T8 derived neurons
**
Ofd1wt
Ofd1ko
C
Relative GLI3R level in Ofd1 knockout mice
***
D
Relative GLI3R level in T8 derived neurons
*
control
SFN
E
DMSO
lactacystin
pCMV-Myc control
pCMV-Myc-BBS4
Flag-NICD
NICD
GAPDH
Relative NICD level
N/A
1.00
0.74
1.88
Relative NICD level
pCMV Myc
pCMV-Myc -BBS4
F
MG132
GFP-JAG1
140
Relative JAG1 level
1.00
0.25
0.40
Relative JAG1 level

Figure S3

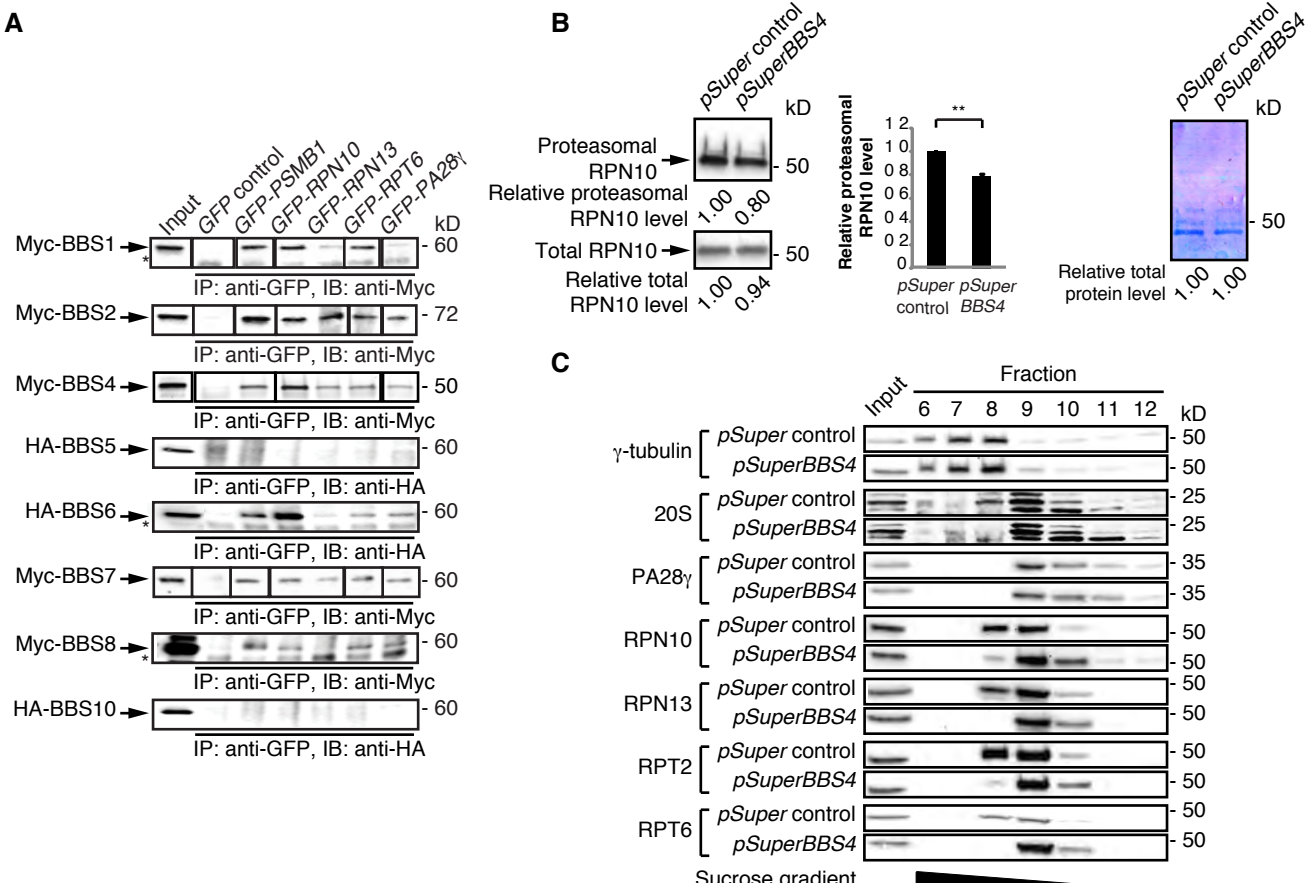
A
Input
GFP control
GFP-PSMB1
GFP-RPN10
GFP-RPN13
GFP-RPT6
GFP-PA28γ
kD
Myc-BBS1
60
IP: anti-GFP, IB: anti-Myc
Myc-BBS2
72
IP: anti-GFP, IB: anti-Myc
Myc-BBS4
50
IP: anti-GFP, IB: anti-Myc
HA-BBS5
60
IP: anti-GFP, IB: anti-HA
HA-BBS6
60
IP: anti-GFP, IB: anti-HA
Myc-BBS7
60
IP: anti-GFP, IB: anti-Myc
Myc-BBS8
60
IP: anti-GFP, IB: anti-Myc
HA-BBS10
60
IP: anti-GFP, IB: anti-HA
B
pSuper control
pSuperBBS4
kD
Proteasomal RPN10
50
Relative proteasomal RPN10 level
1.00
0.80
Total RPN10
50
Relative total RPN10 level
1.00
0.94
Relative proteasomal RPN10 level
1.2
1.0
0.8
0.6
0.4
0.2
0
**
pSuper control
pSuper BBS4
pSuper control
pSuperBBS4
kD
50
Relative total protein level
1.00
1.00
C
Input
Fraction
6 7 8 9 10 11 12
kD
γ-tubulin
pSuper control
pSuperBBS4
50
50
20S
pSuper control
pSuperBBS4
25
25
PA28γ
pSuper control
pSuperBBS4
35
35
RPN10
pSuper control
pSuperBBS4
50
50
RPN13
pSuper control
pSuperBBS4
50
50
RPT2
pSuper control
pSuperBBS4
50
50
RPT6
pSuper control
pSuperBBS4
50
50
Sucrose gradient

Figure S4

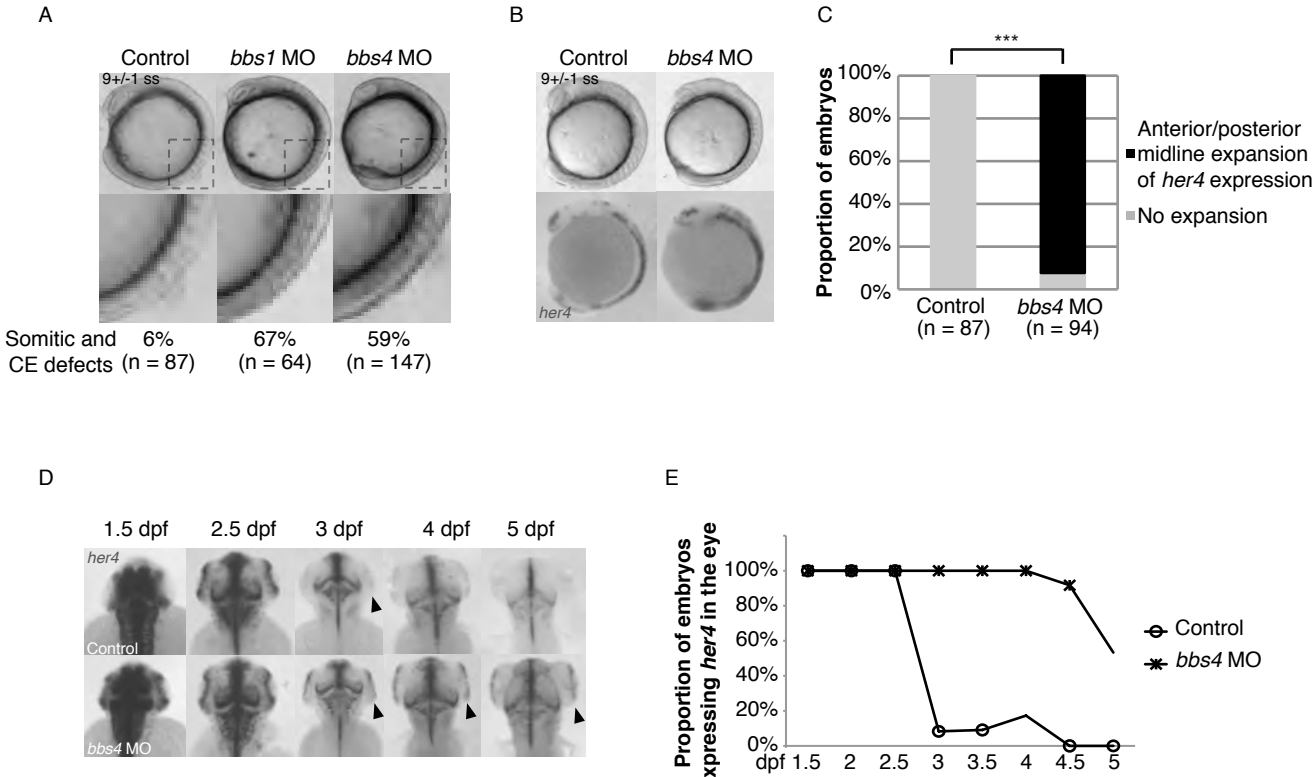
A
Control
bbs1 MO
bbs4 MO
9+/-1 ss
Somitic and CE defects
6% (n = 87)
67% (n = 64)
59% (n = 147)
B
Control
bbs4 MO
9+/-1 ss
her4
C
***
Proportion of embryos
100%
80%
60%
40%
20%
0%
Control (n = 87)
bbs4 MO (n = 94)
Anterior/posterior midline expansion of her4 expression
No expansion
D
1.5 dpf
2.5 dpf
3 dpf
4 dpf
5 dpf
her4
Control
bbs4 MO
E
Proportion of embryos expressing her4 in the eye
100%
80%
60%
40%
20%
0%
dpf 1.5 2 2.5 3 3.5 4 4.5 5
Control
bbs4 MO

Figure S5

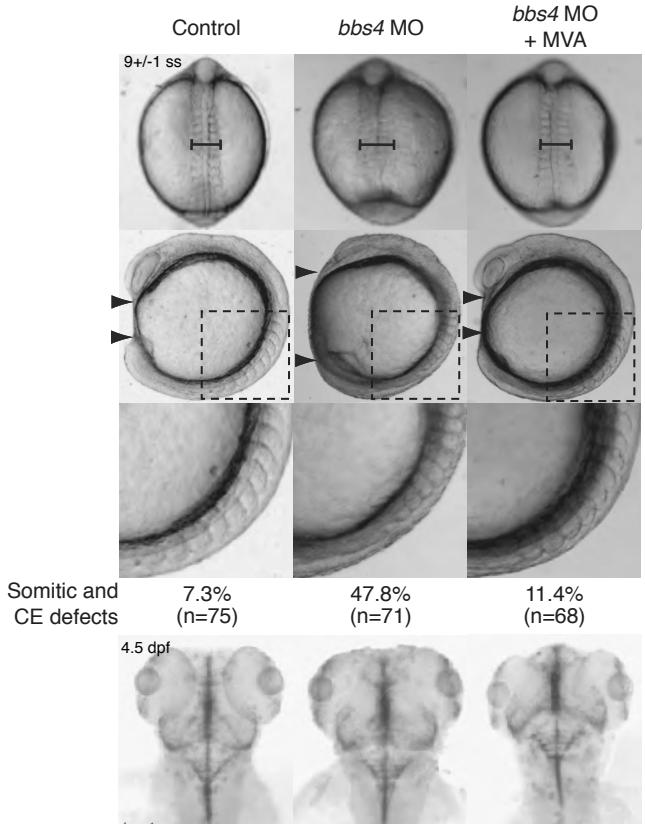
Control
bbs4 MO
bbs4 MO + MVA
9+/-1 ss
Somitic and CE defects
7.3% (n=75)
47.8% (n=71)
11.4% (n=68)
4.5 dpf
her4

Figure S6