Figure 1A

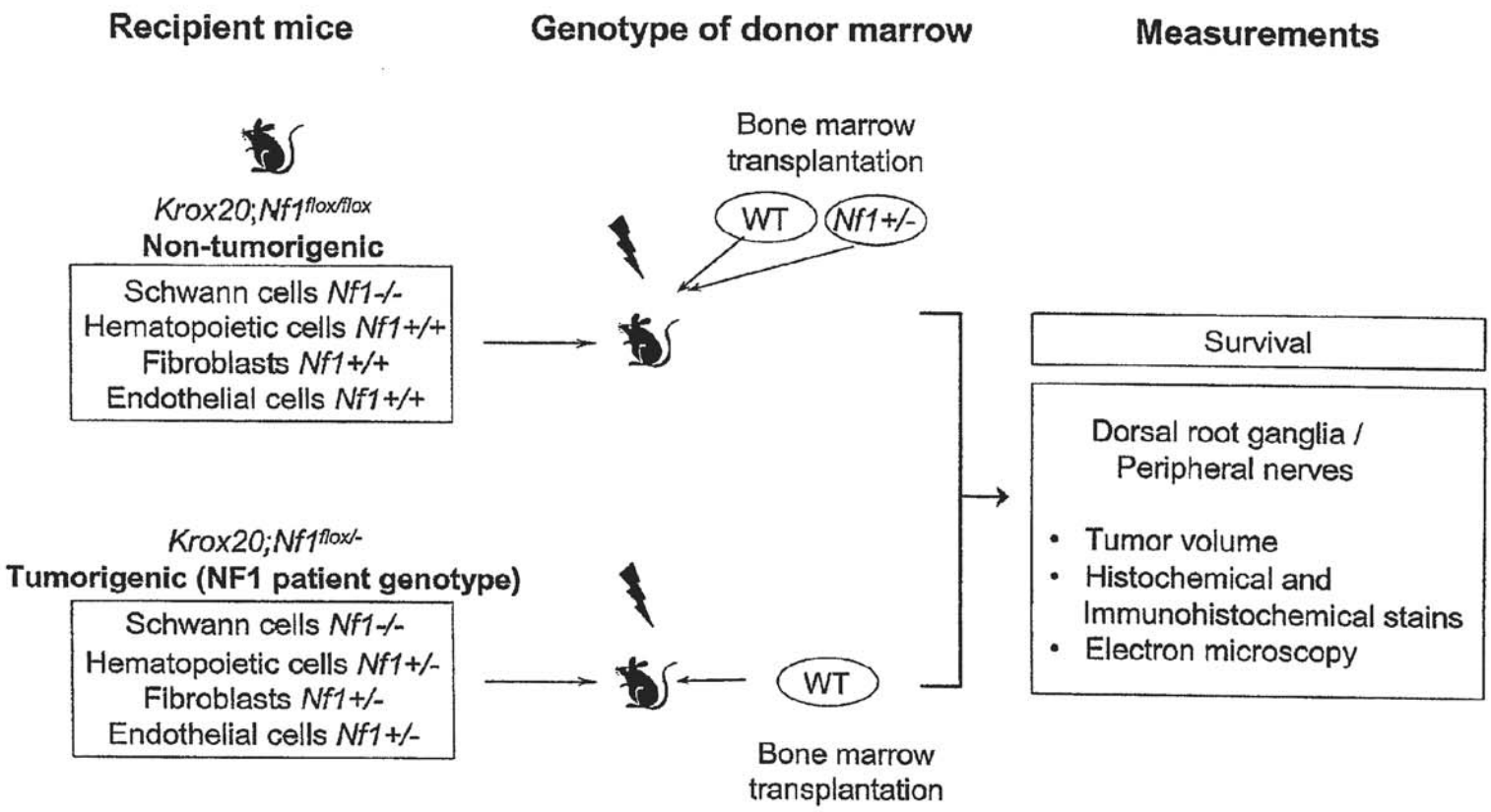
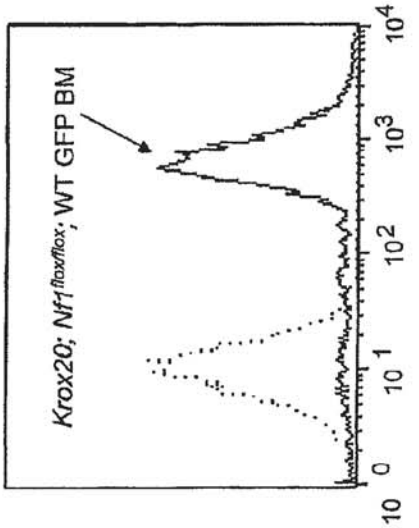
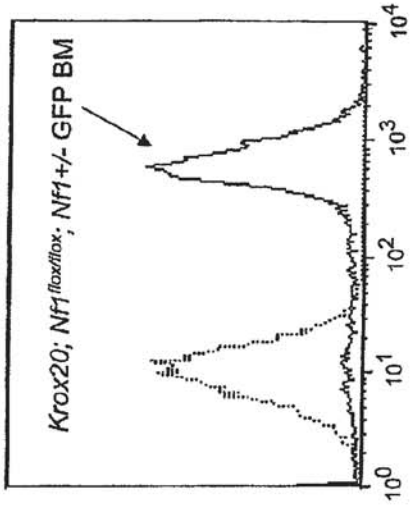

Recipient mice
Genotype of donor marrow
Measurements
Bone marrow transplantation
WT
Nf1+/-
Krox20;Nf1flox/flox
Non-tumorigenic
Schwann cells Nf1-/-
Hematopoietic cells Nf1+/+
Fibroblasts Nf1+/+
Endothelial cells Nf1+/+
Krox20;Nf1flox/-
Tumorigenic (NF1 patient genotype)
Schwann cells Nf1-/-
Hematopoietic cells Nf1+/-
Fibroblasts Nf1+/-
Endothelial cells Nf1+/-
WT
Bone marrow transplantation
Survival
Dorsal root ganglia / Peripheral nerves
• Tumor volume
• Histochemical and Immunohistochemical stains
• Electron microscopy

Figure 1 B


Krox20; Nf1flox/flox; WT GFP BM
10 0
10 1
10 2
10 3
10 4


Krox20; Nf1flox/flox; Nf1+/- GFP BM
10 0
10 1
10 2
10 3
10 4

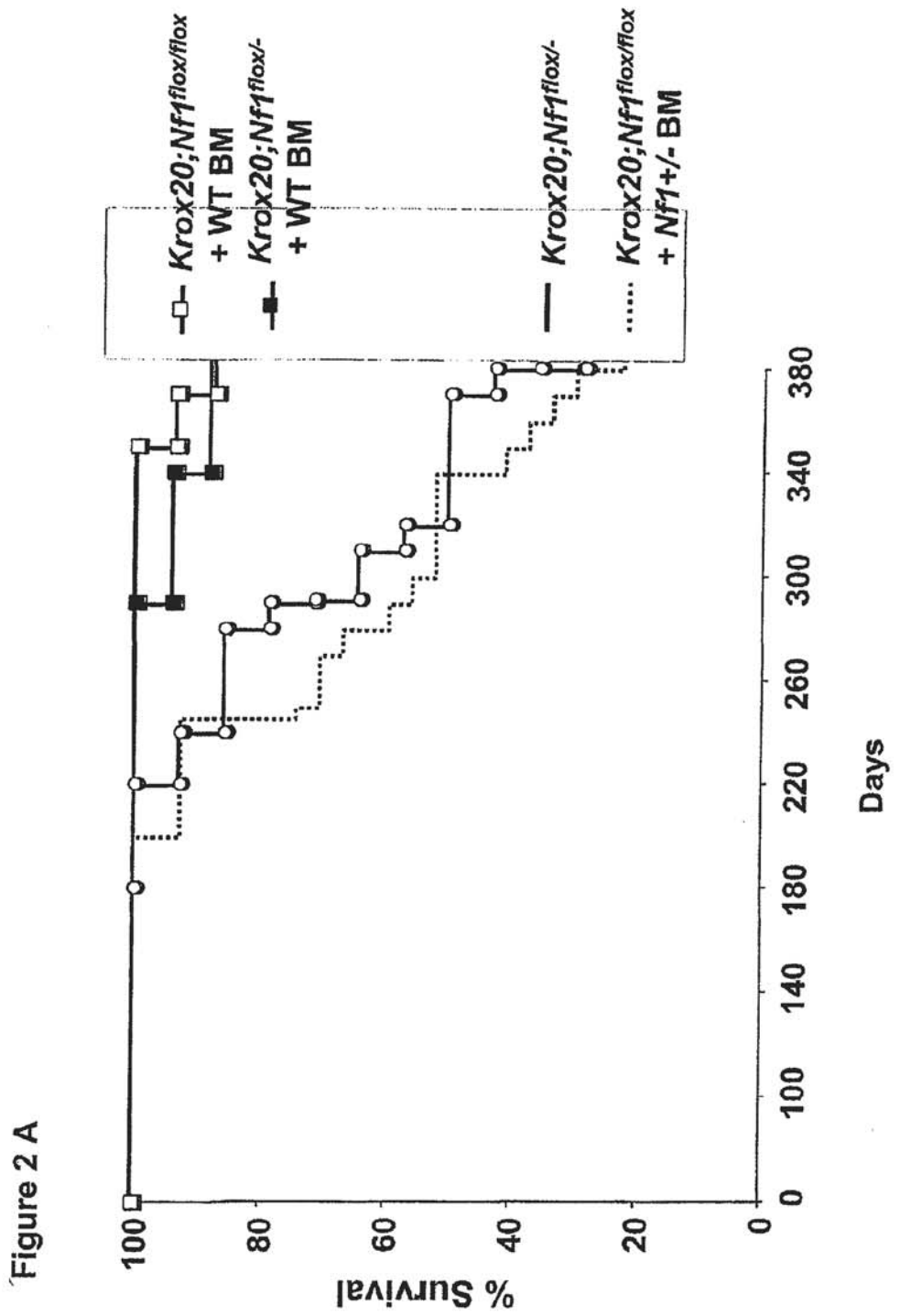

Krox20;Nf1flox/flox + WT BM
Krox20;Nf1flox/- + WT BM
Krox20;Nf1flox/-
Krox20;Nf1flox/flox + Nf1+/- BM
% Survival
100
80
60
40
20
0
0
100
140
180
220
260
300
340
380
Days

Figure 2 A

Figure 2 B

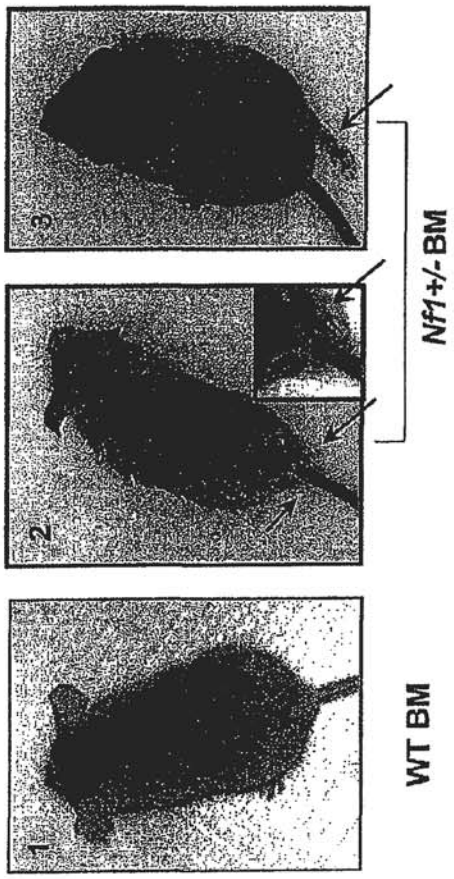
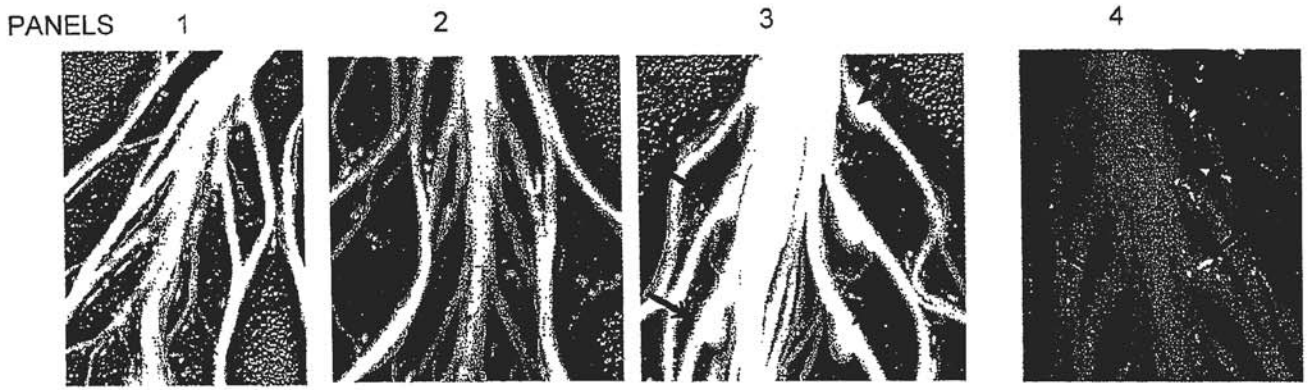


Figure 2 C.



WT bone marrow *Nf1* +/- bone marrow

Krox20; Nf1floxIflox

Figure 3 A

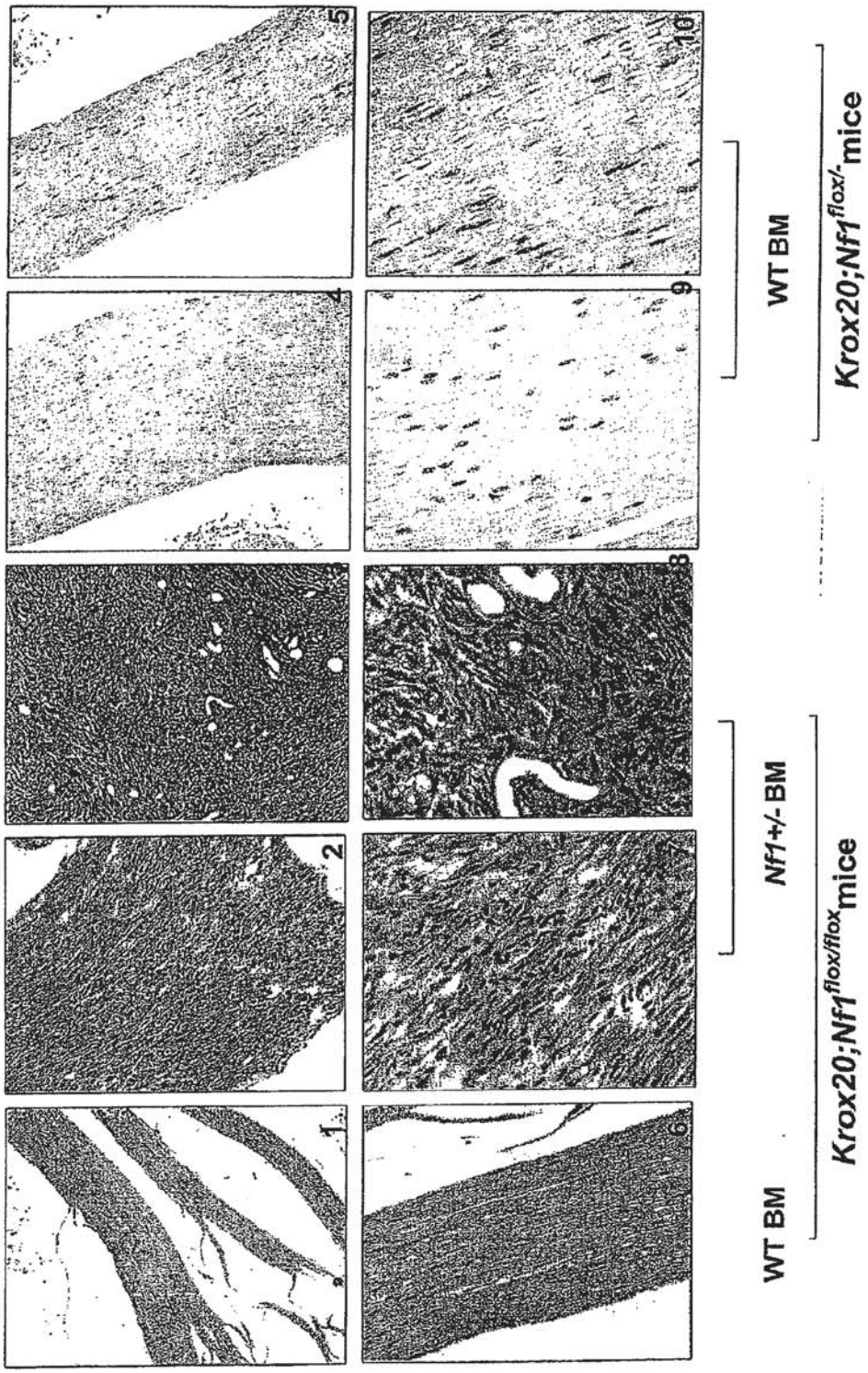

1
2
3
4
5
6
7
8
9
10
WT BM
Nf1+/- BM
Krox20;Nf1flox/flox mice
WT BM
Krox20;Nf1flox/- mice

Figure 3 B

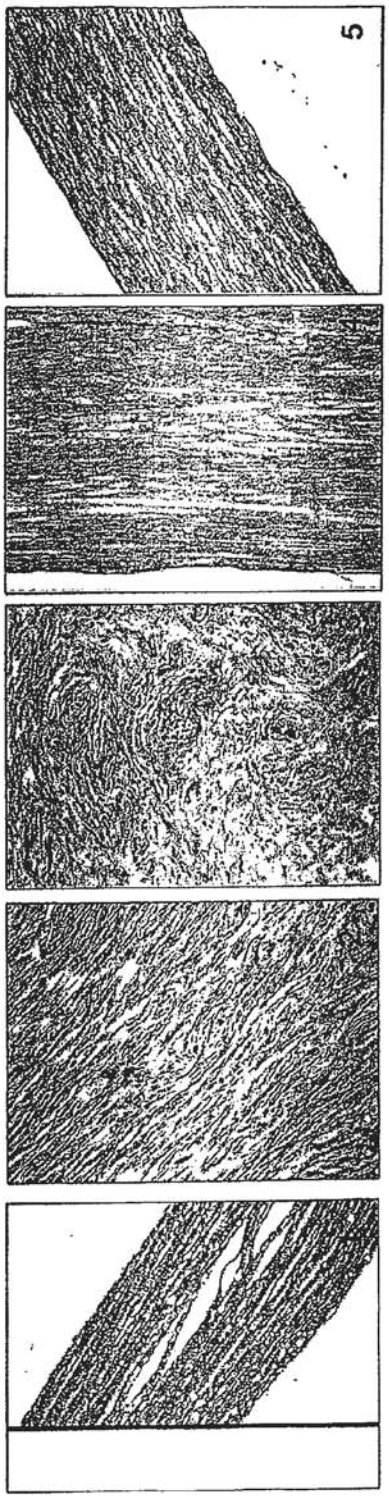

5
WT BM
Nf1+/- BM
WT BM
Krox20;Nf1flox/flox mice
Krox20;Nf1flox/- mice

Figure 3 C

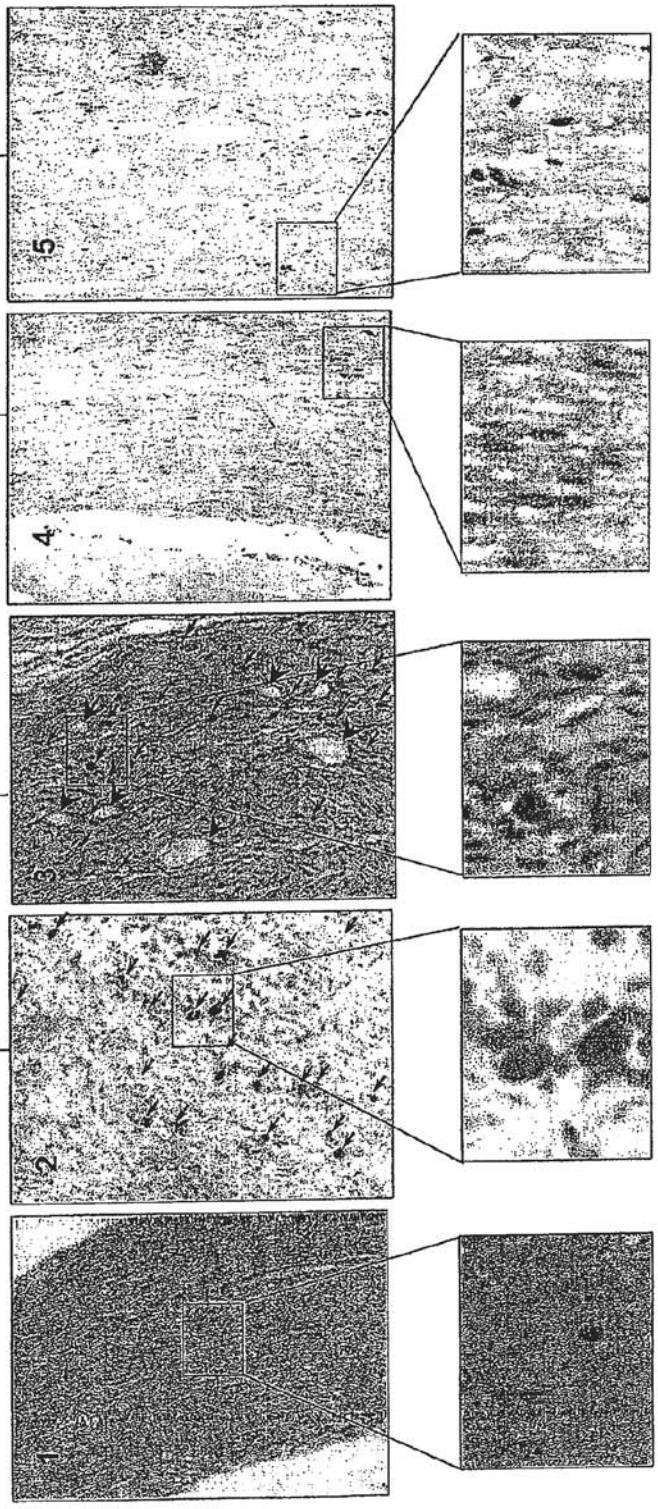

Krox20;Nf1flox/flox mice
WT BM
Nf1+/- BM
Krox20;Nf1flox/- mice
WT BM
1
2
3
4
5

Figure 3 D

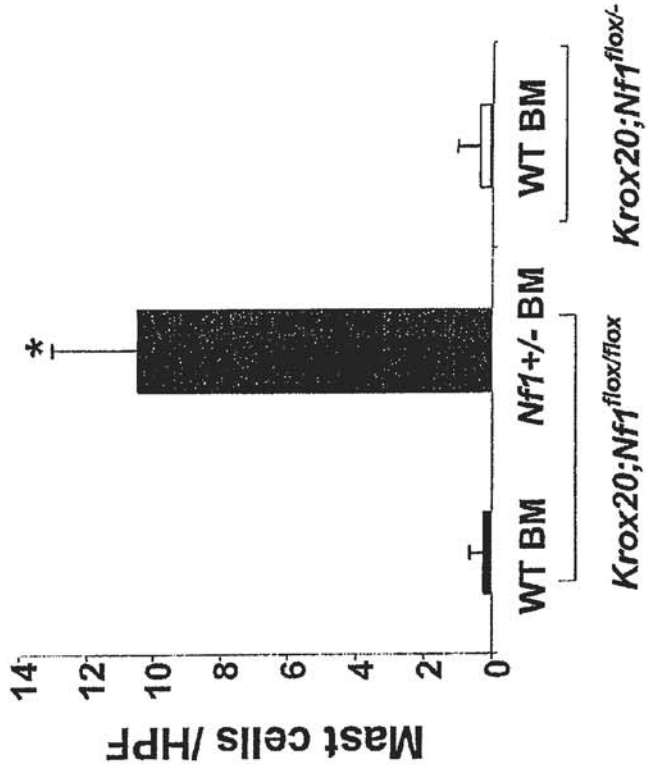
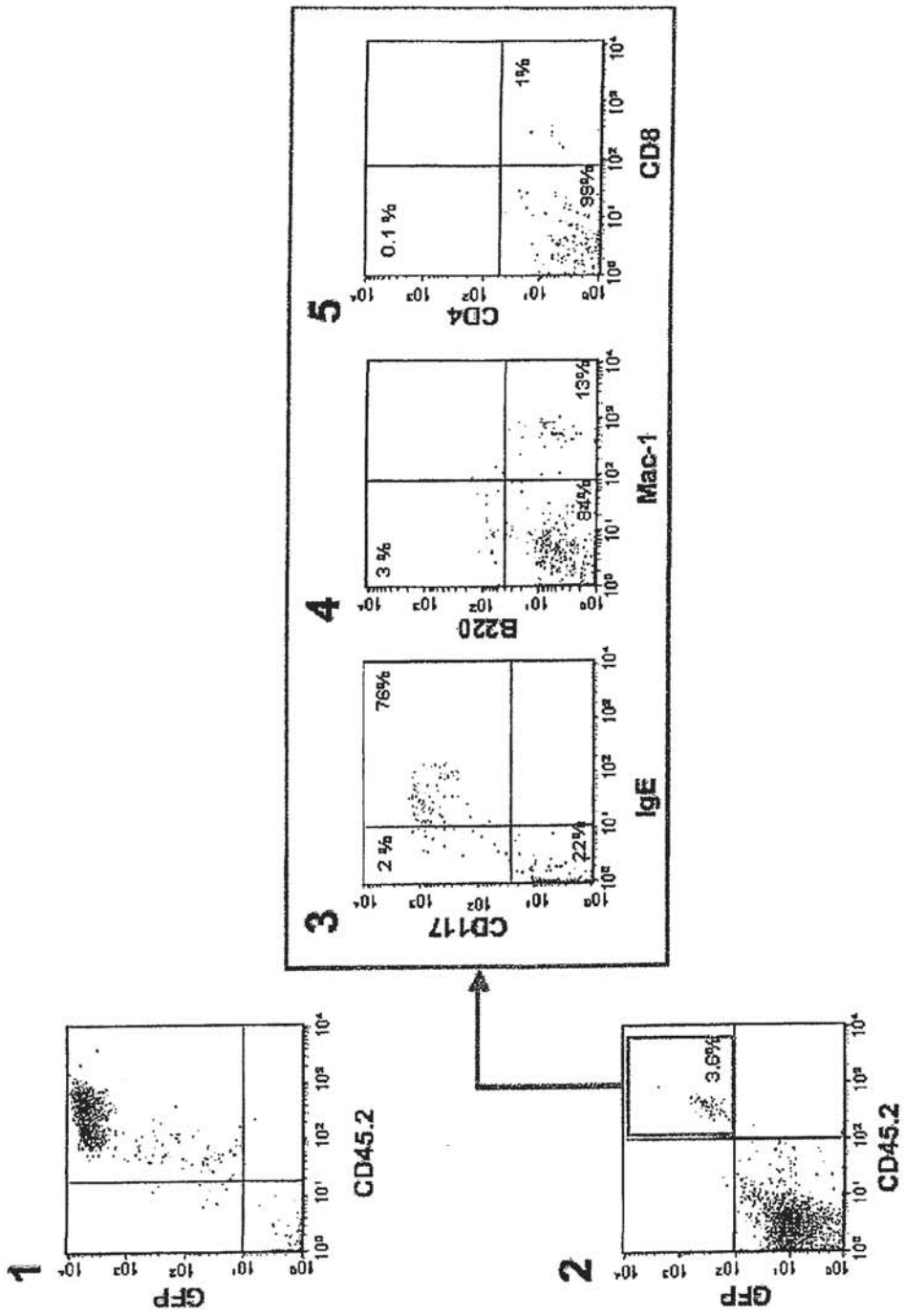

Mast cells /HPF
14
12
10
8
6
4
2
0
*
WT BM
Nf1+/- BM
WT BM
Krox20;Nf1flox/flox
Krox20;Nf1flox/-

Figure 3E


1
GFP
CD45.2
2
GFP
CD45.2
3.6%
3
CD117
IgE
2 %
76%
22%
4
B220
Mac-1
3 %
84%
13%
5
CD4
CD8
0.1 %
99%
1%

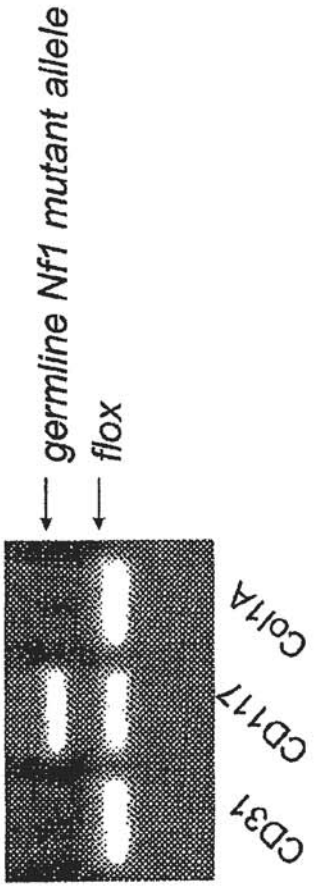


Figure 3F

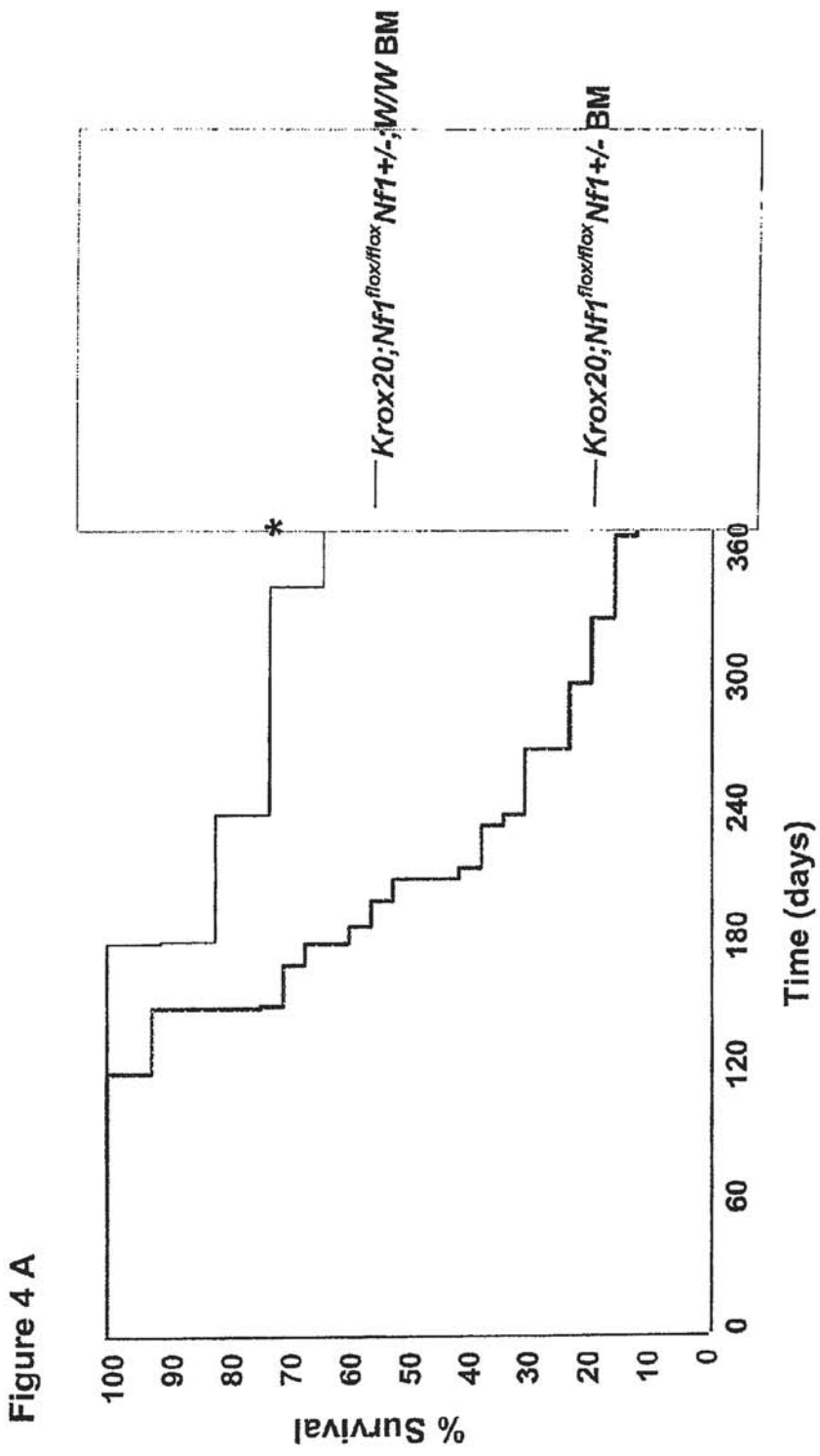

Figure 4 A
% Survival
100
90
80
70
60
50
40
30
20
10
0
0
60
120
180
240
300
360
Time (days)
*
—Krox20;Nf1flox/flox Nf1+/-;W/W BM
—Krox20;Nf1flox/flox Nf1+/- BM

Figure 4 B





Nf1+/- BM

Nf1+/-W^v/W^v BM

Krox20;Nf1$^{flox/flox}$

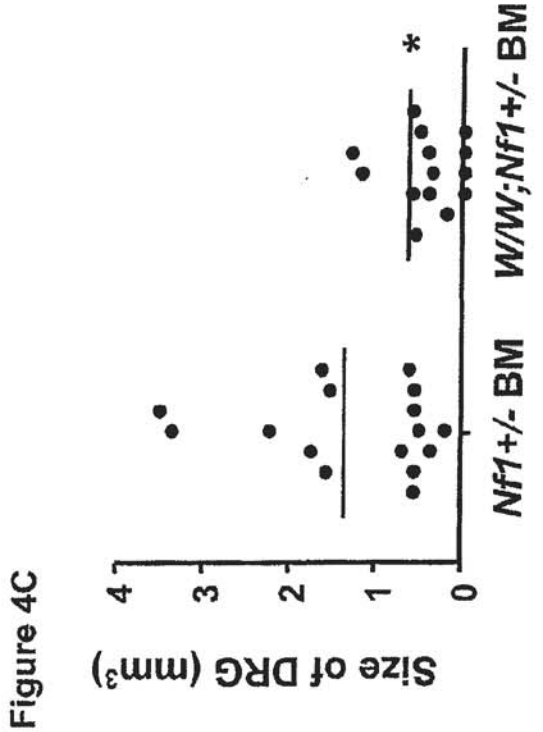


Figure 4C

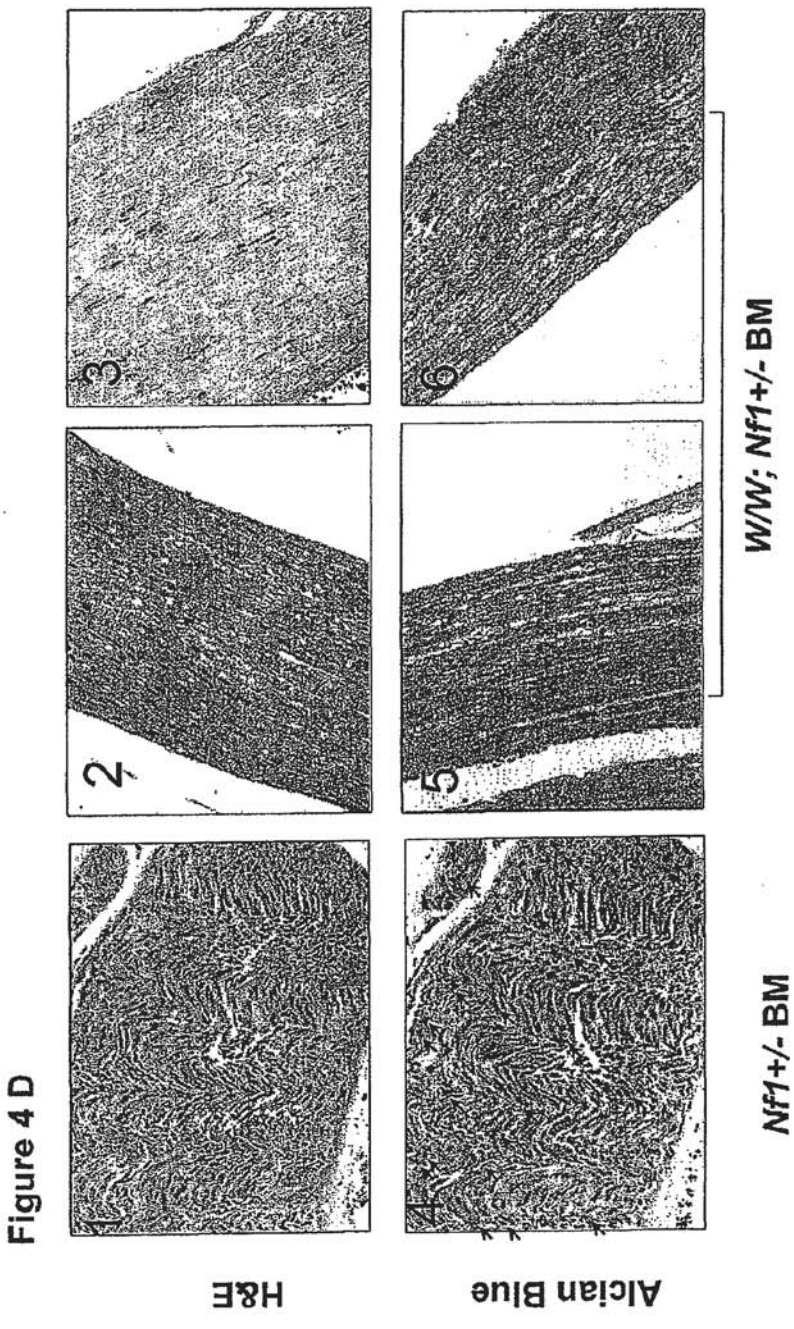
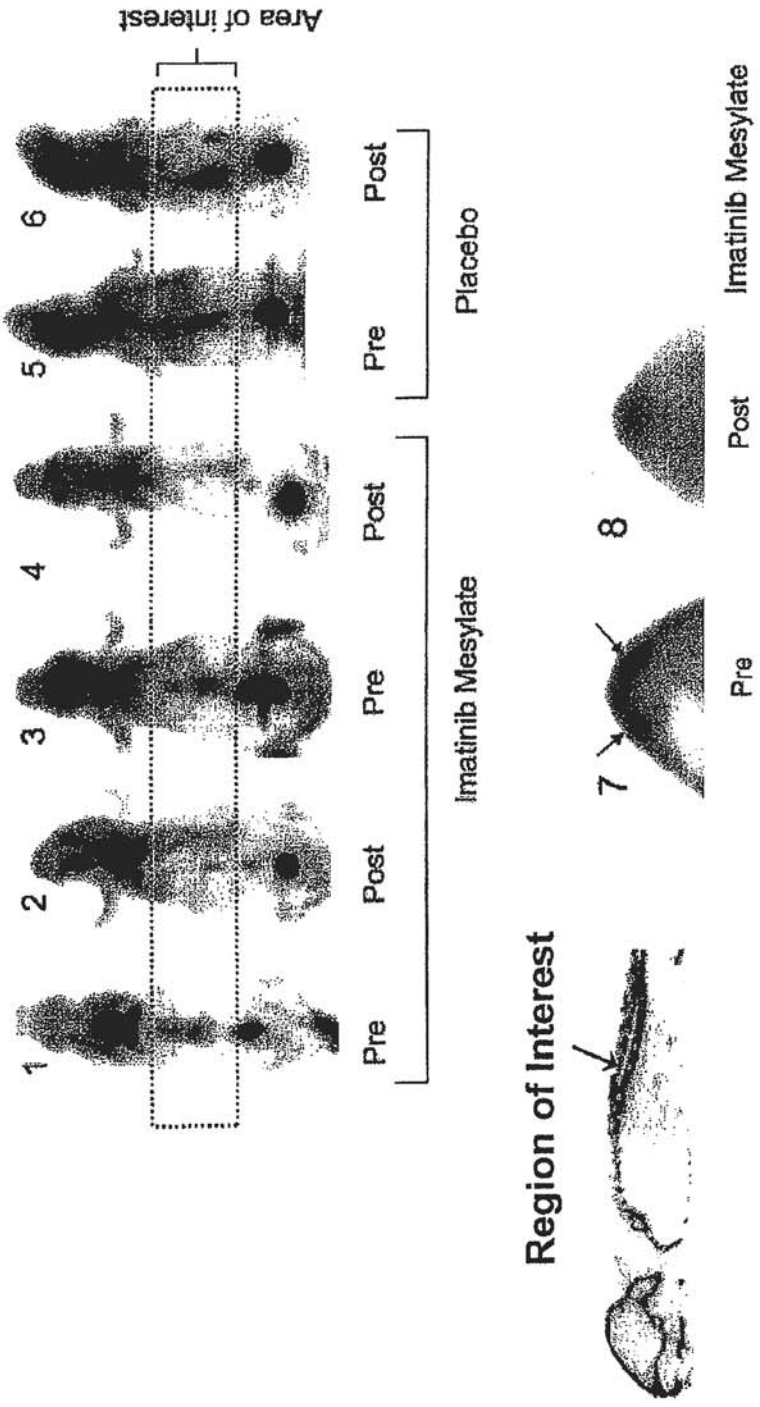

Figure 4 D
H&E
Alcian Blue
Nf1+/- BM
W/W; Nf1+/- BM
2
3
5
6

Figure 5 A


Area of interest
1
2
3
4
5
6
Pre
Post
Pre
Post
Pre
Post
Imatinib Mesylate
Placebo
Region of Interest
7
8
Pre
Post
Imatinib Mesylate

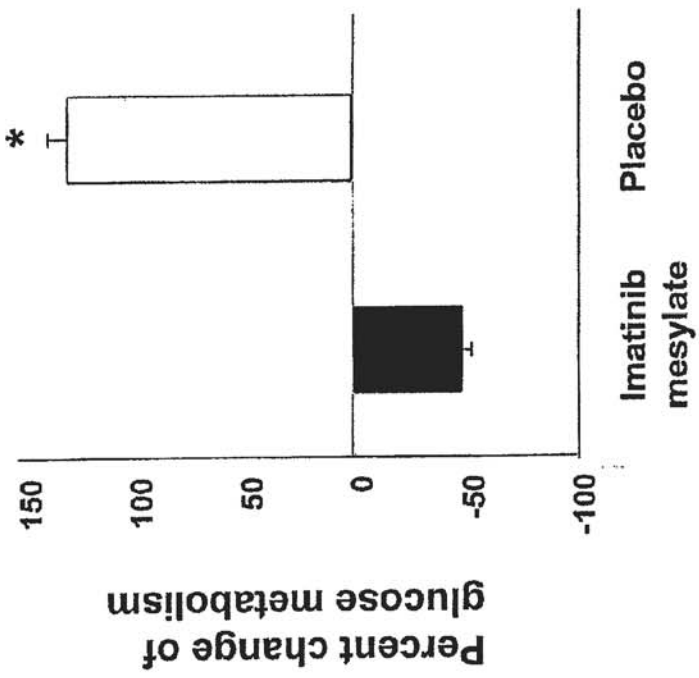

Percent change of glucose metabolism
150
100
50
0
-50
-100
*
Imatinib mesylate
Placebo

Figure 5B .

Figure 5 C

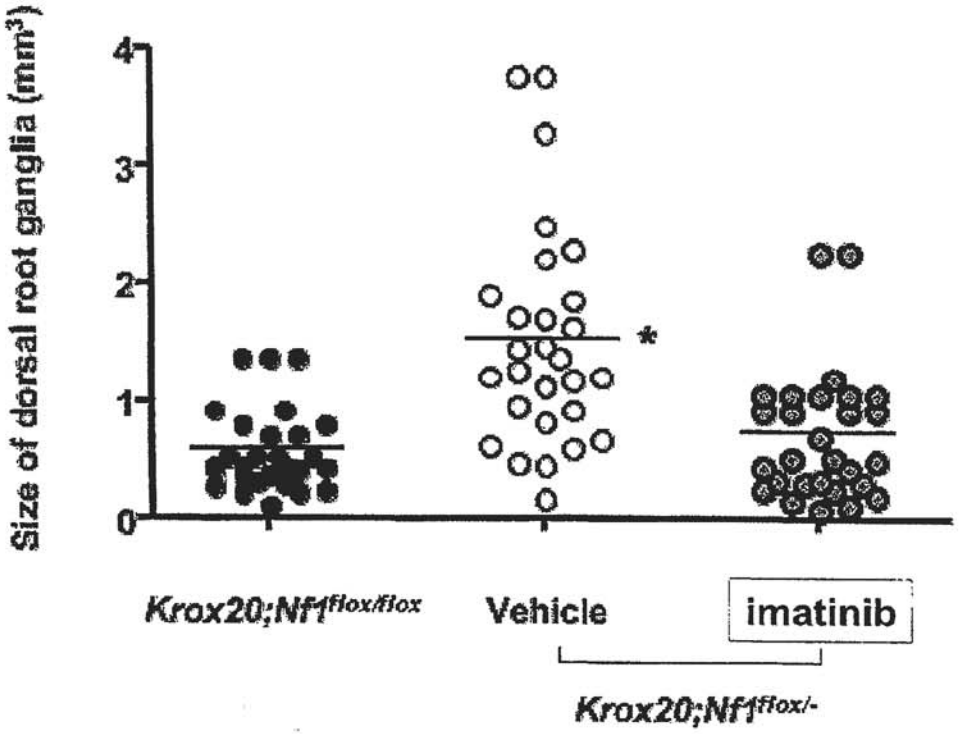

Size of dorsal root ganglia (mm3)
4
3
2
1
0
*
Krox20;Nf1flox/flox
Vehicle
imatinib
Krox20;Nf1flox/-

Figure 5 D

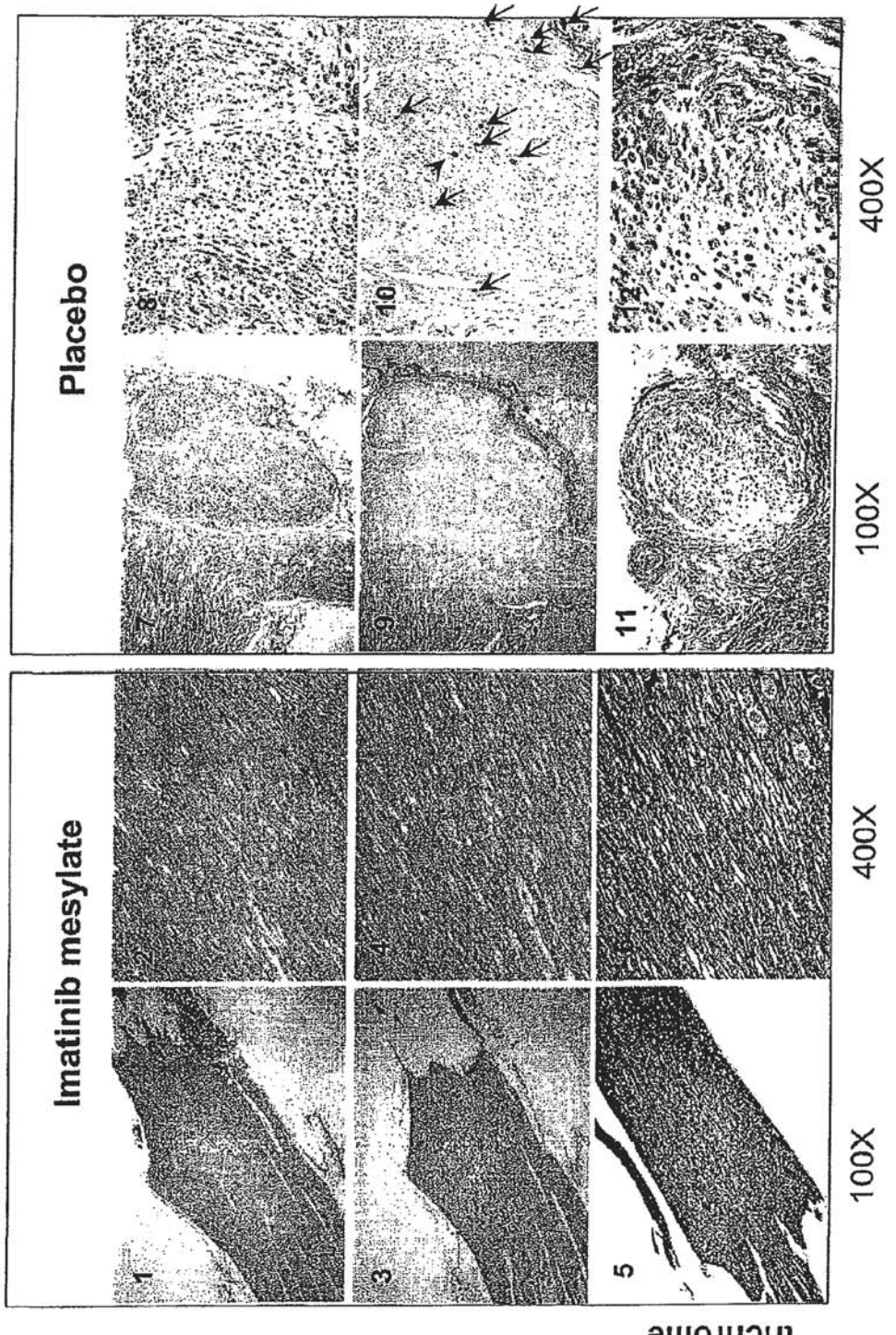
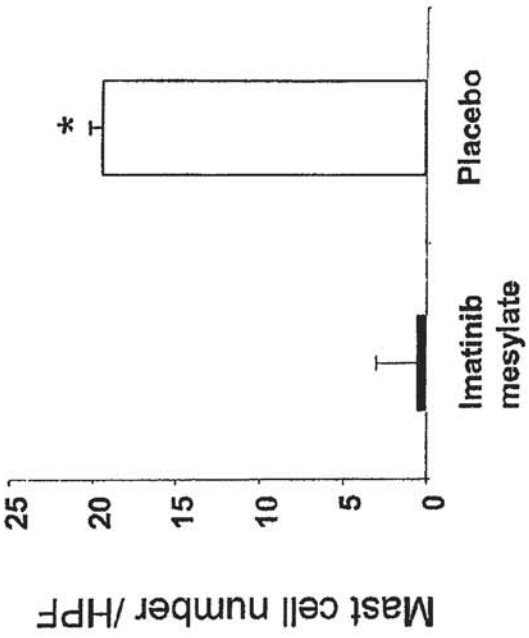

Imatinib mesylate
Placebo
H & E
Alcian Blue
Masson's trichrome
100X
400X
100X
400X
1
2
3
4
5
6
7
8
9
10
11
12

Figure 5E


Mast cell number /HPF
25
20
15
10
5
0
*
Imatinib mesylate
Placebo

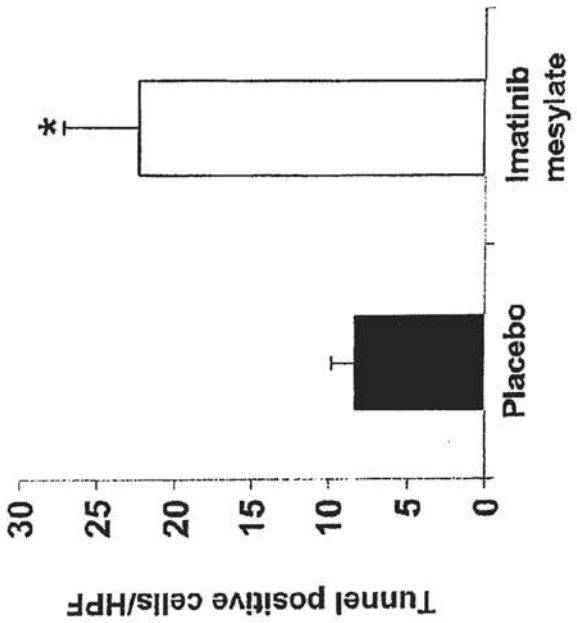
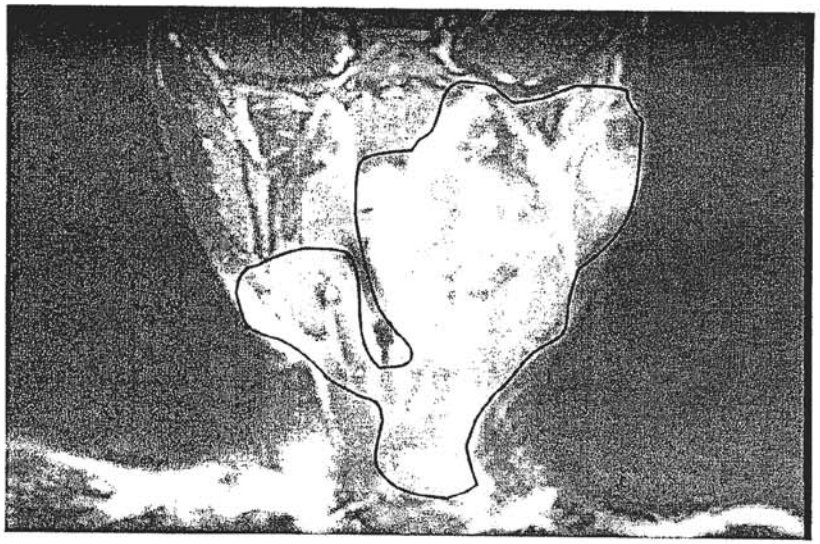
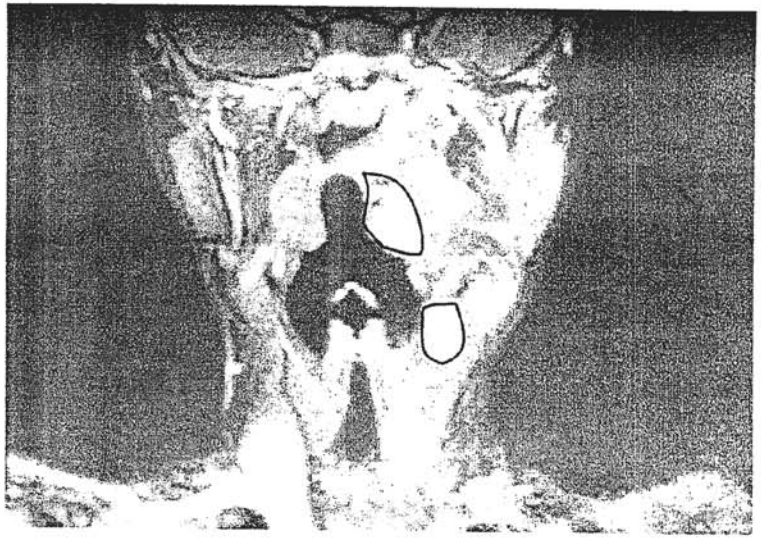

Tunnel positive cells/HPF
30
25
20
15
10
5
0
*
Placebo
Imatinib mesylate

Figure 5F

Figure 6



Before



Three months treatment with imatinib mesylate

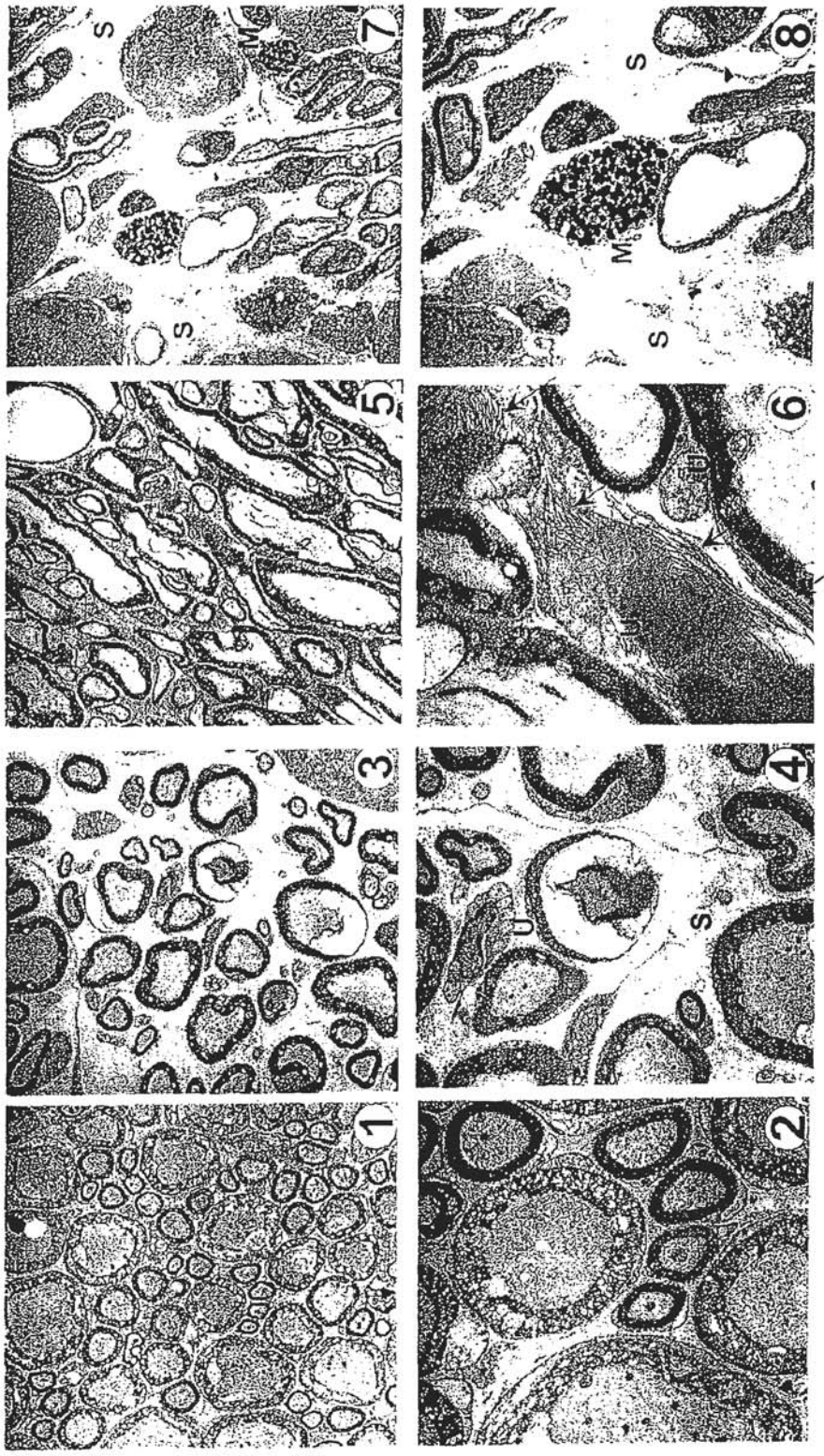
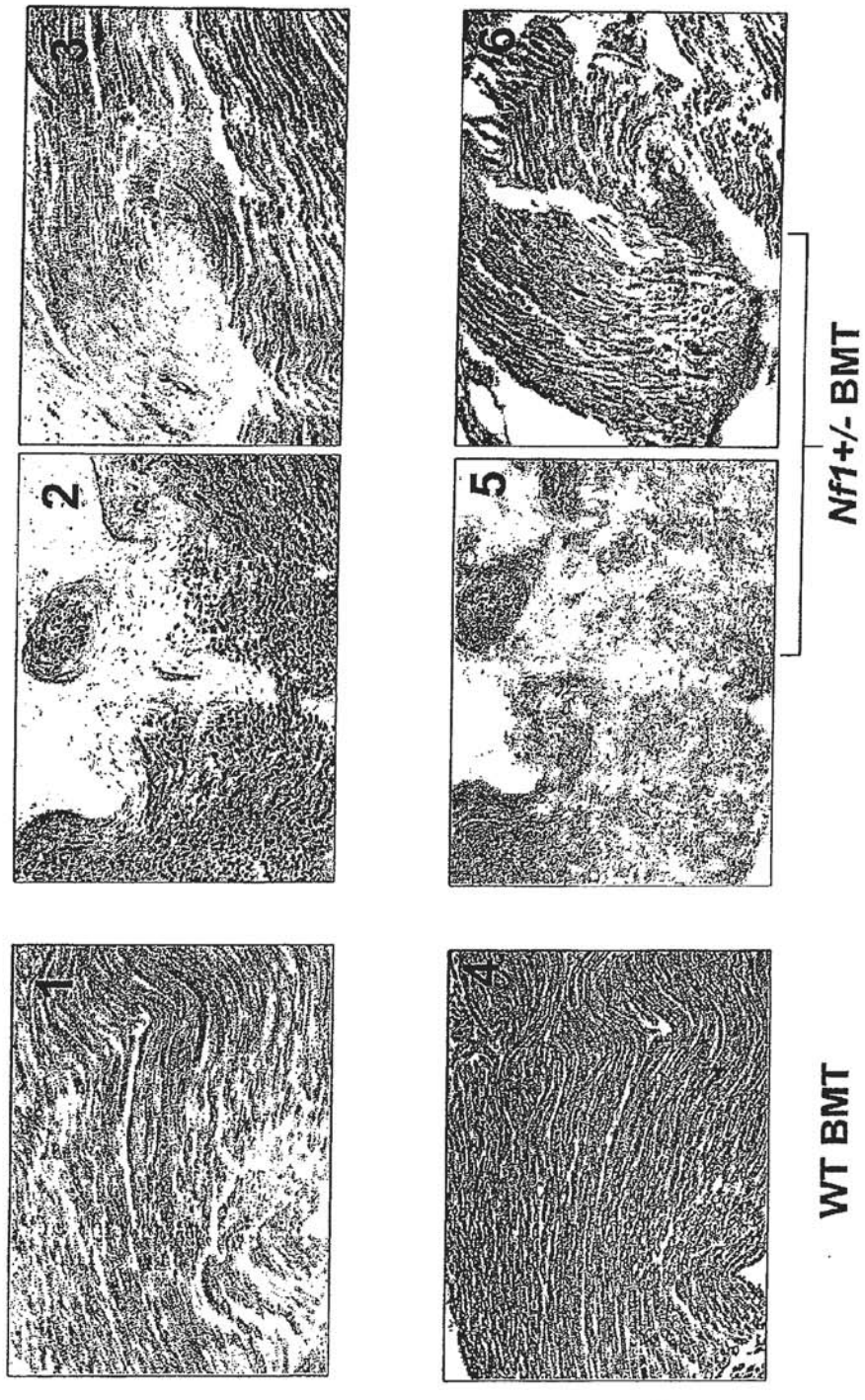


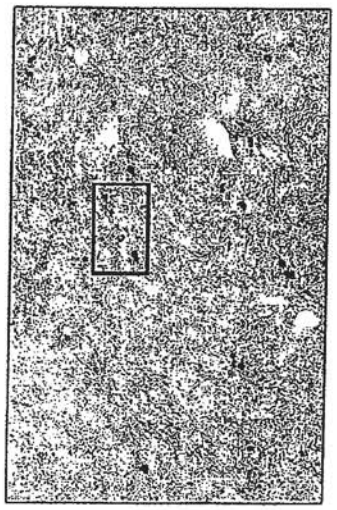
Figure 7

Figure 8





600X



100X

Toludine blue

Figure 9

Figure 10

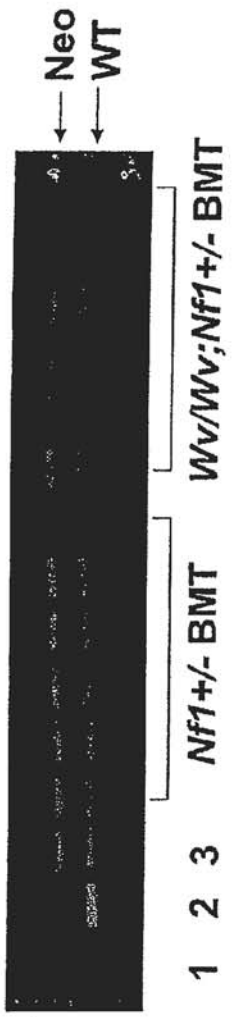
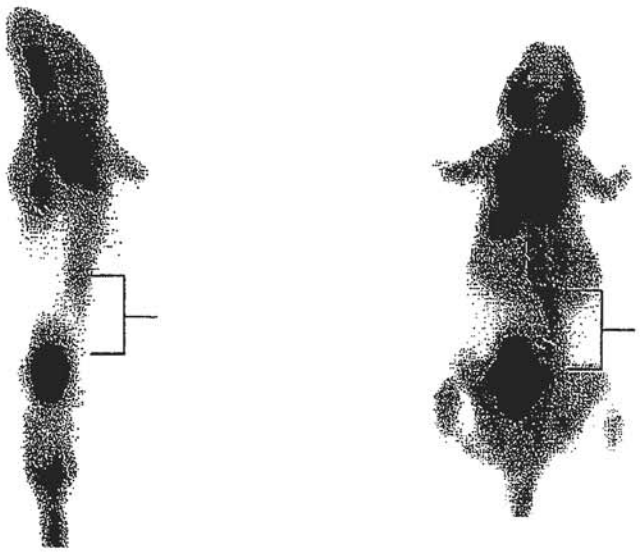


Figure 11A



Krox20;Nf1$^{flox/flox}$ ***Krox20;Nf1$^{flox/-}$***

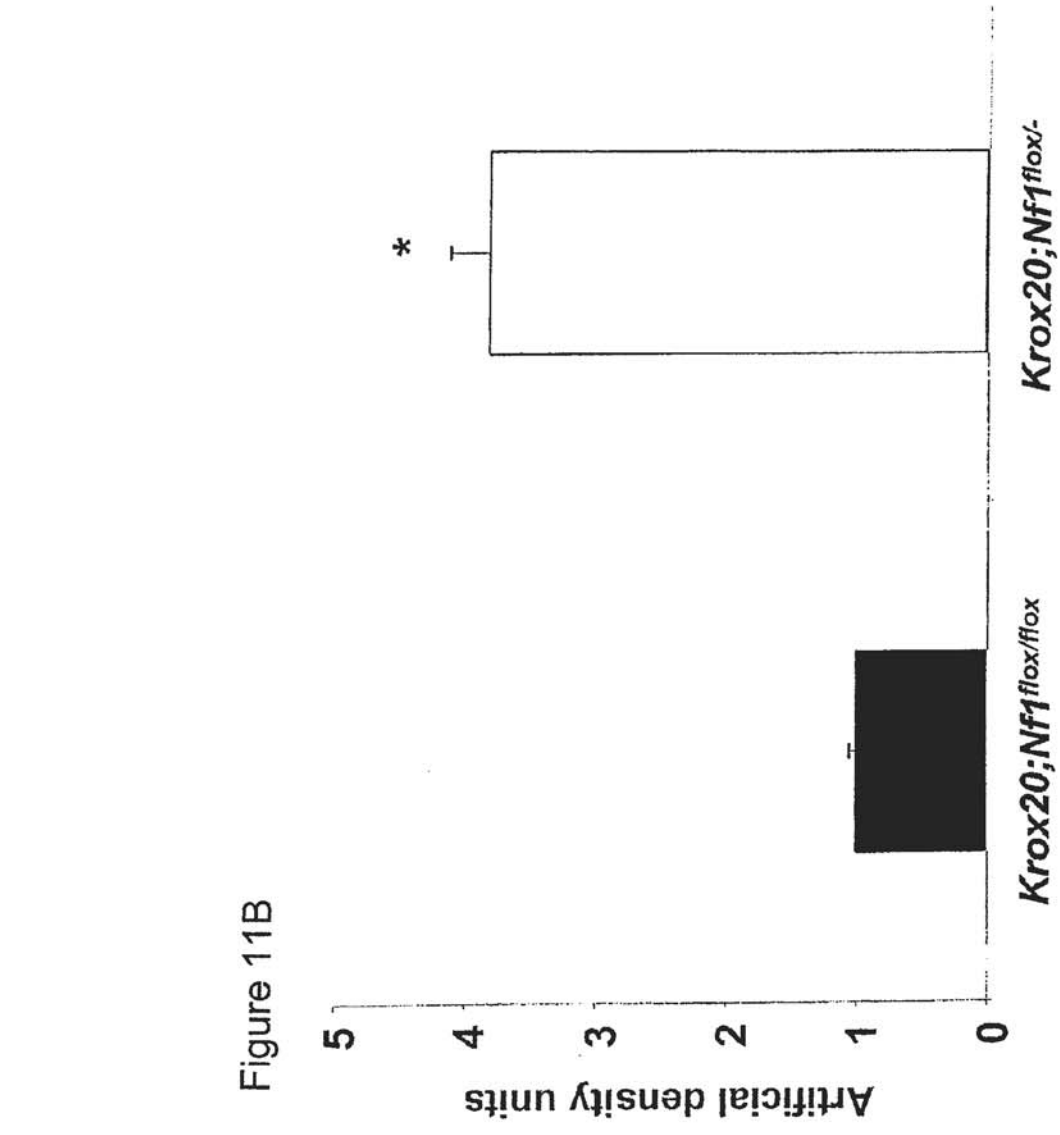
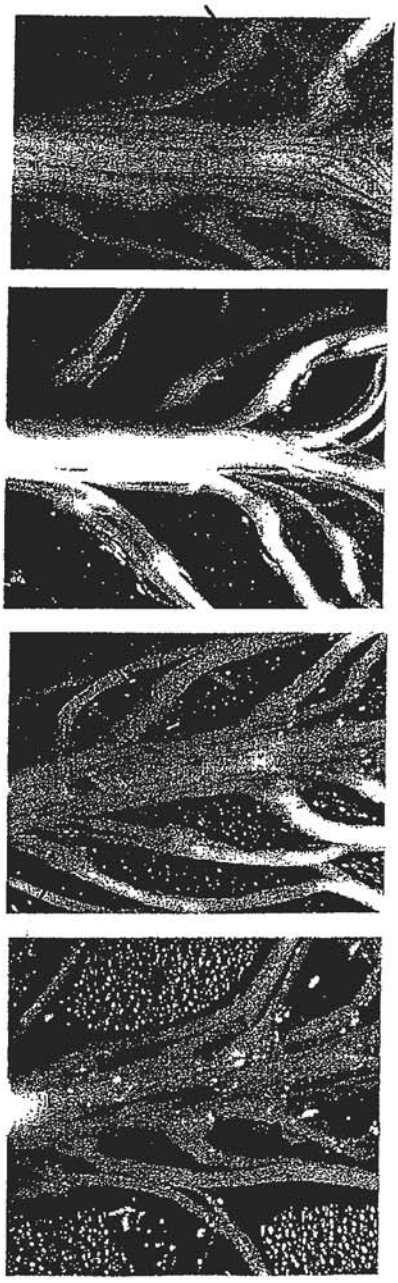

Artificial density units
5
4
3
2
1
0
*
Krox20;Nf1flox/flox
Krox20;Nf1flox/-

Figure 11B

Figure 11C



Krox20;Nf1$^{flox/flox}$

Krox20;Nf1$^{flox/-}$

Figure 12

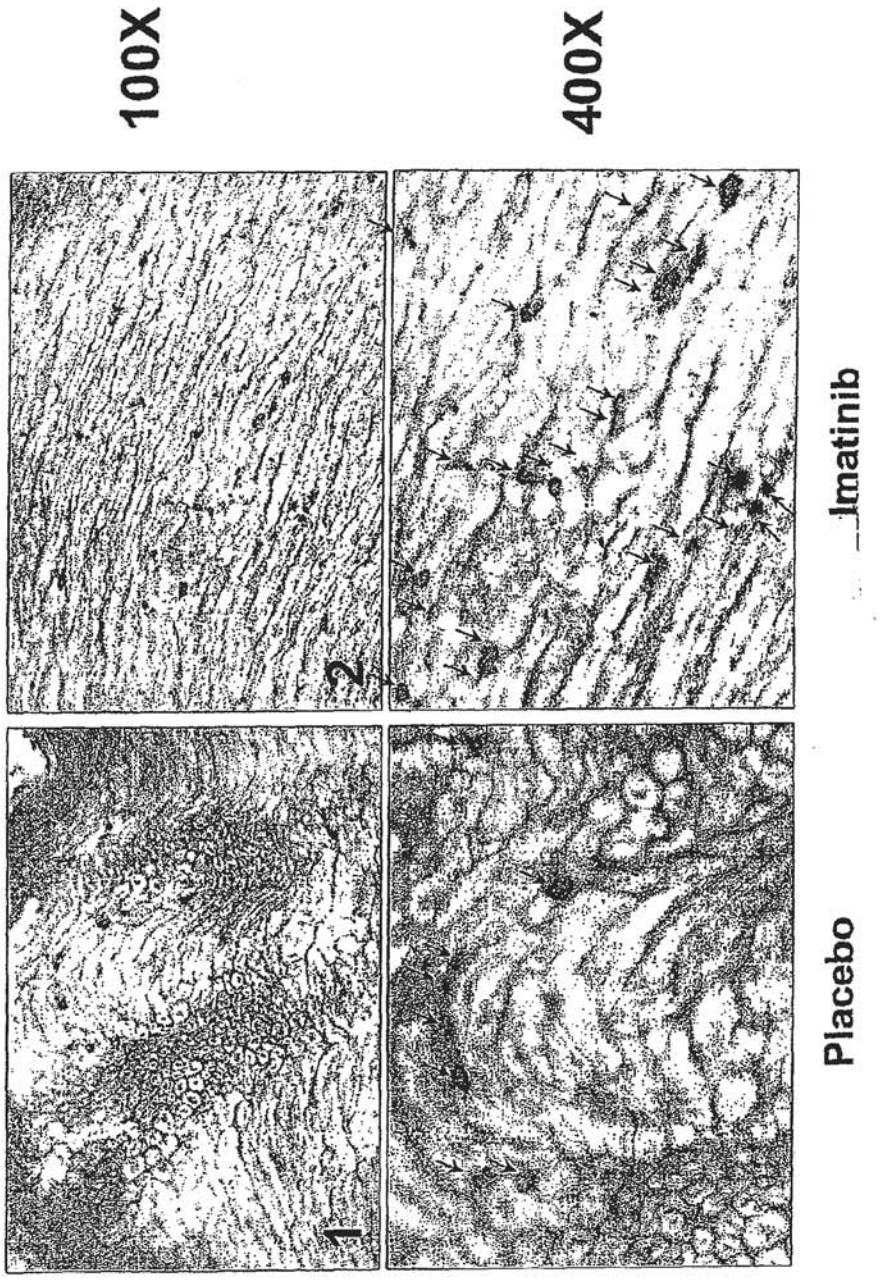


Figure 13A

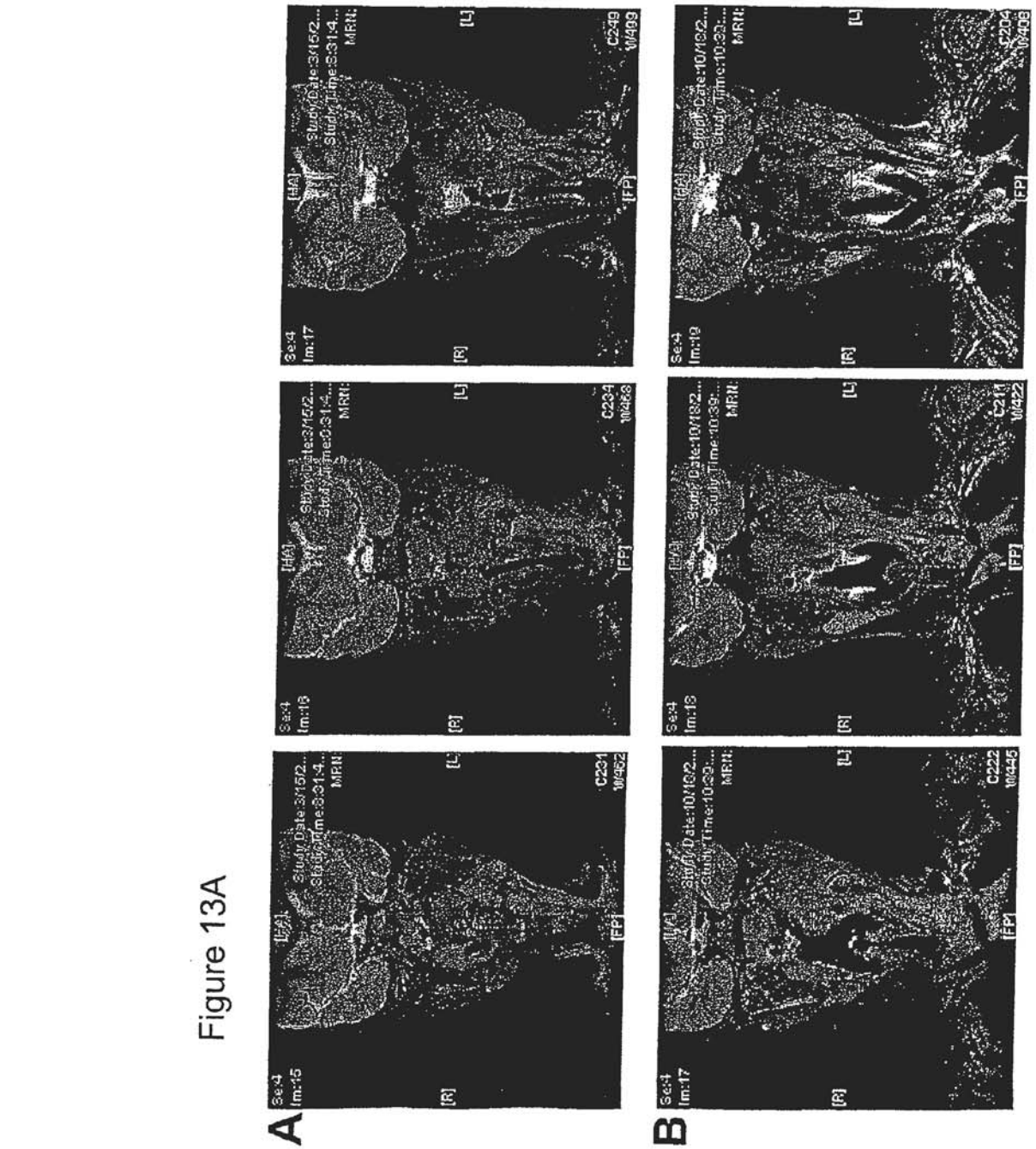
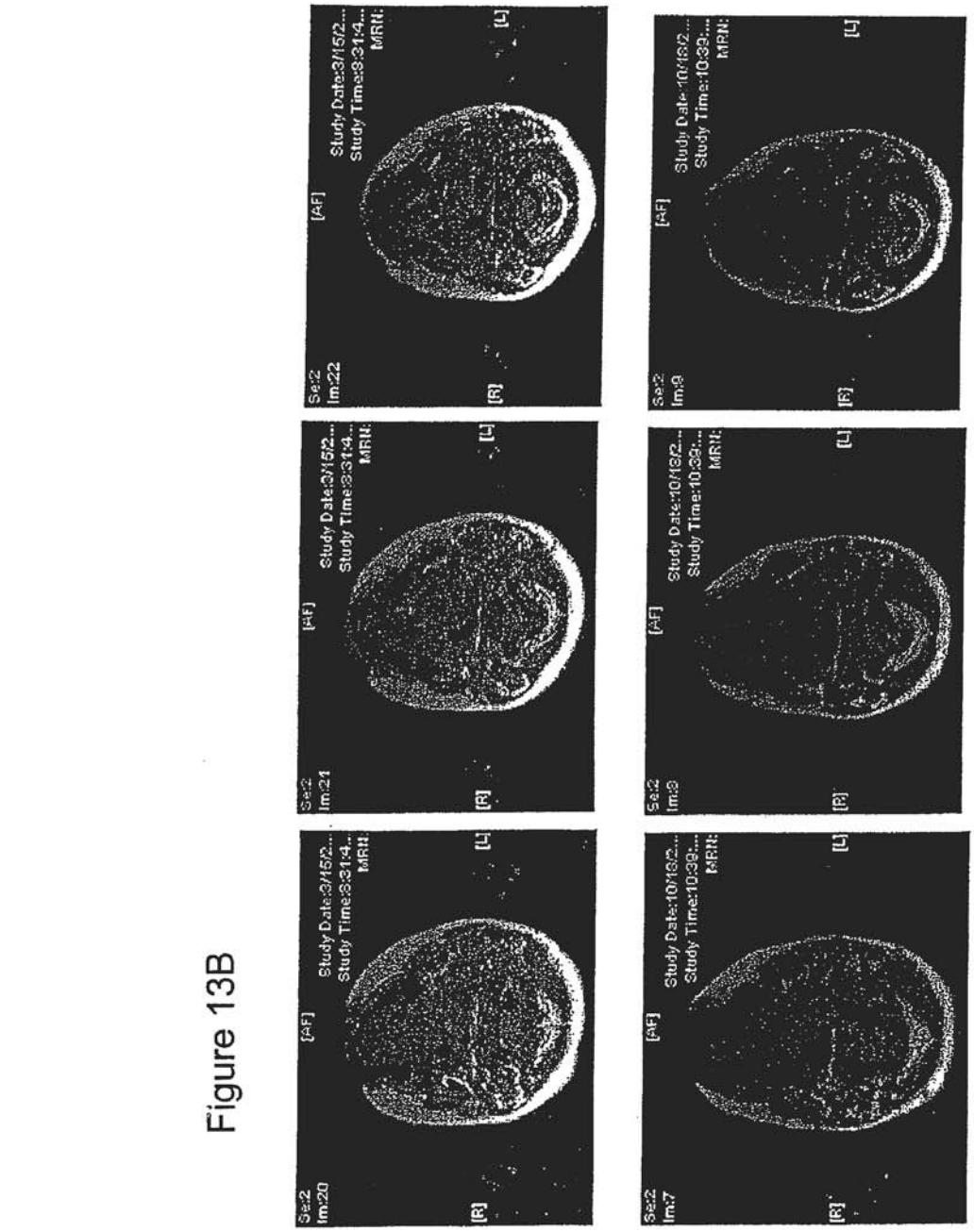

A
B
Ser:4
Im:15
Ser:4
Im:16
Ser:4
Im:17
Ser:4
Im:18
Ser:4
Im:19
[HA]
[FP]
[R]
[L]
C231
W462
C234
W468
C249
W499
C222
W445
C211
W422
C204
W408

Figure 13B


C
D
[AF]
[R]
[L]
Se:2
Im:20
Im:21
Im:22
Im:7
Im:8
Im:9
Study Date:3/15/2...
Study Time:8:31:4...
Study Date:10/18/2...
Study Time:10:39:...
MRN: