

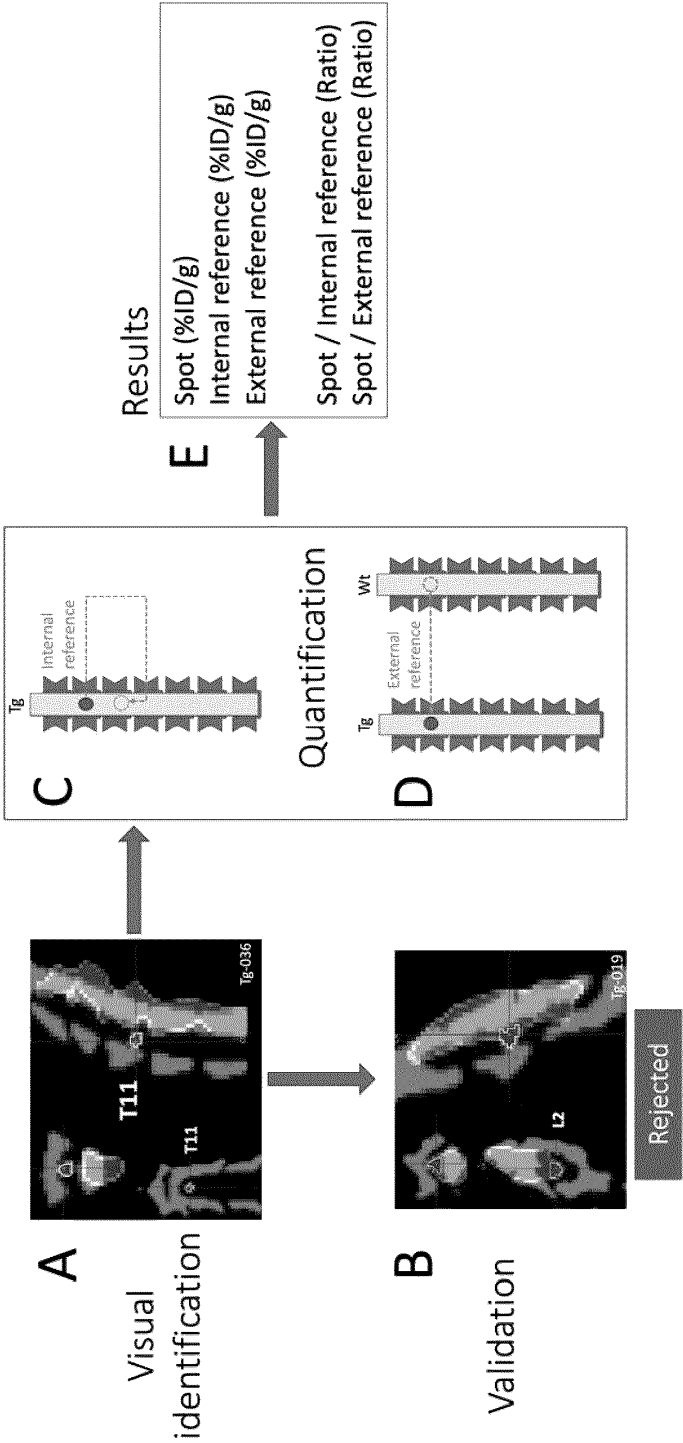


Fig. 1

F

ALS mice AP101 d10	Age 91d	Age 110d	Age 126d
Number of spots	3	5	14
spots/mouse	0.6	1.25	1.6
mice w. spots/total mice	2/5 (40%)	4/4 (100%)	6/9 (67%)
Average intensity [%ID/g]	9.5	7.9	8.0

G

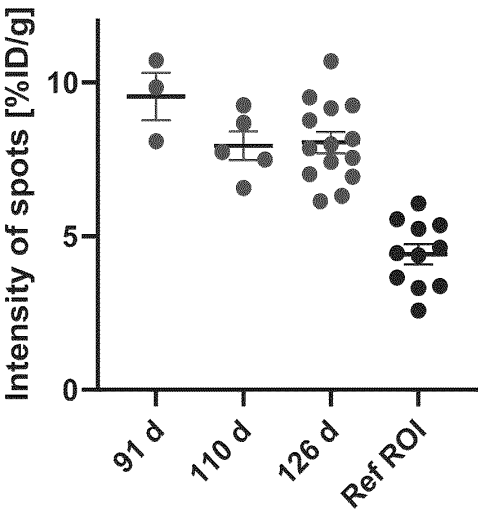
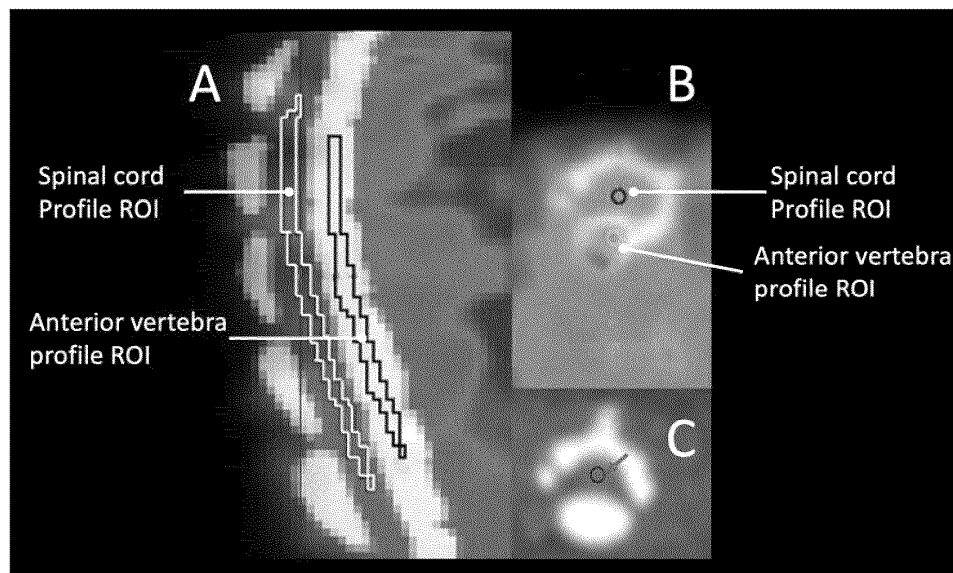


Fig. 1 (continued)

**Fig. 2**

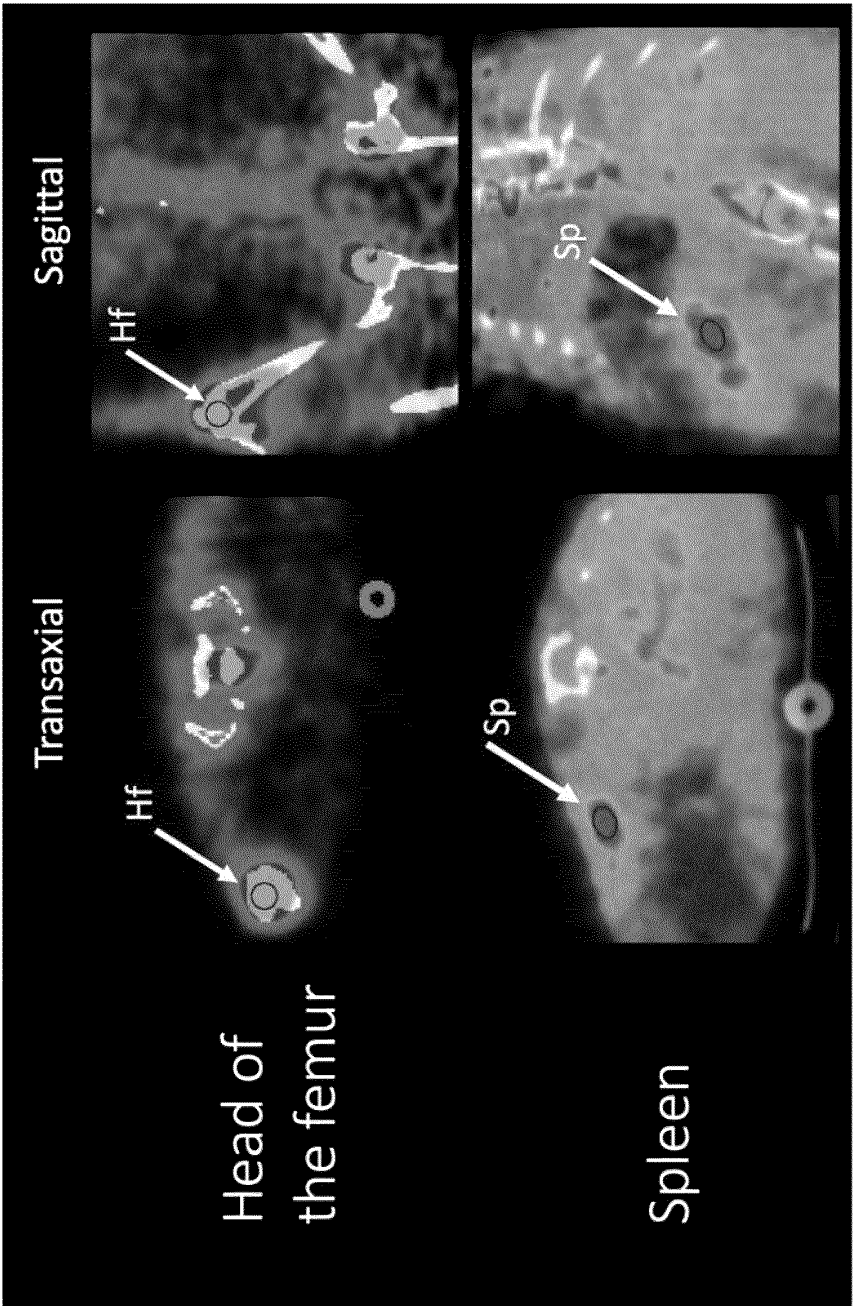


Fig. 3

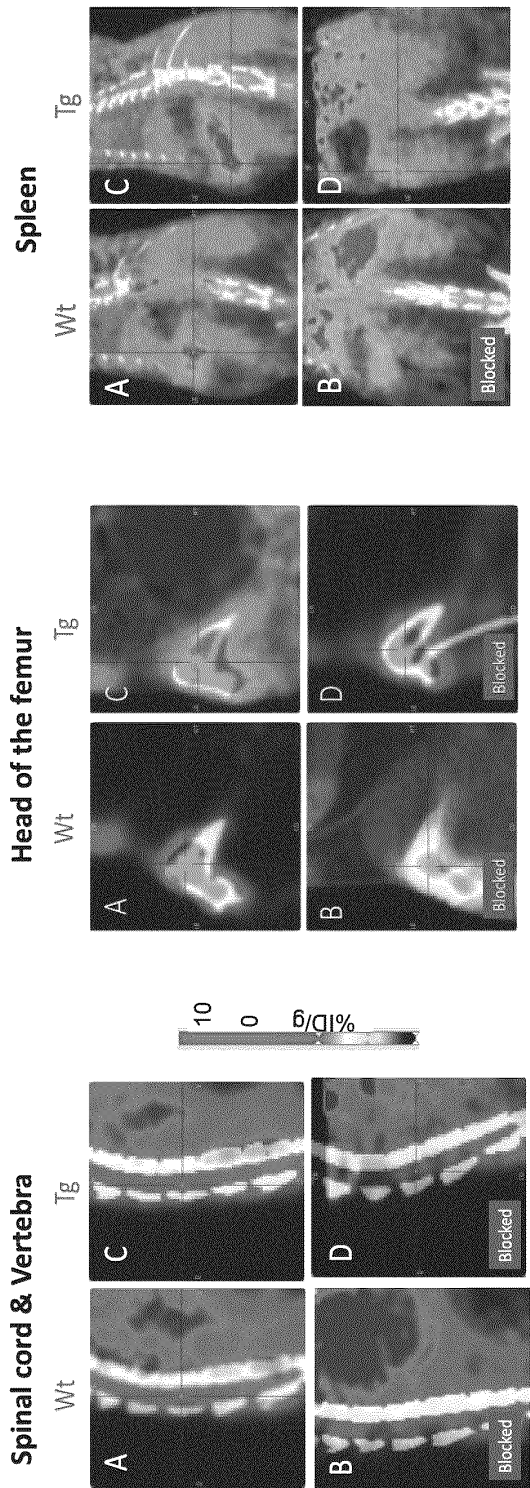


Fig. 3 (continued)

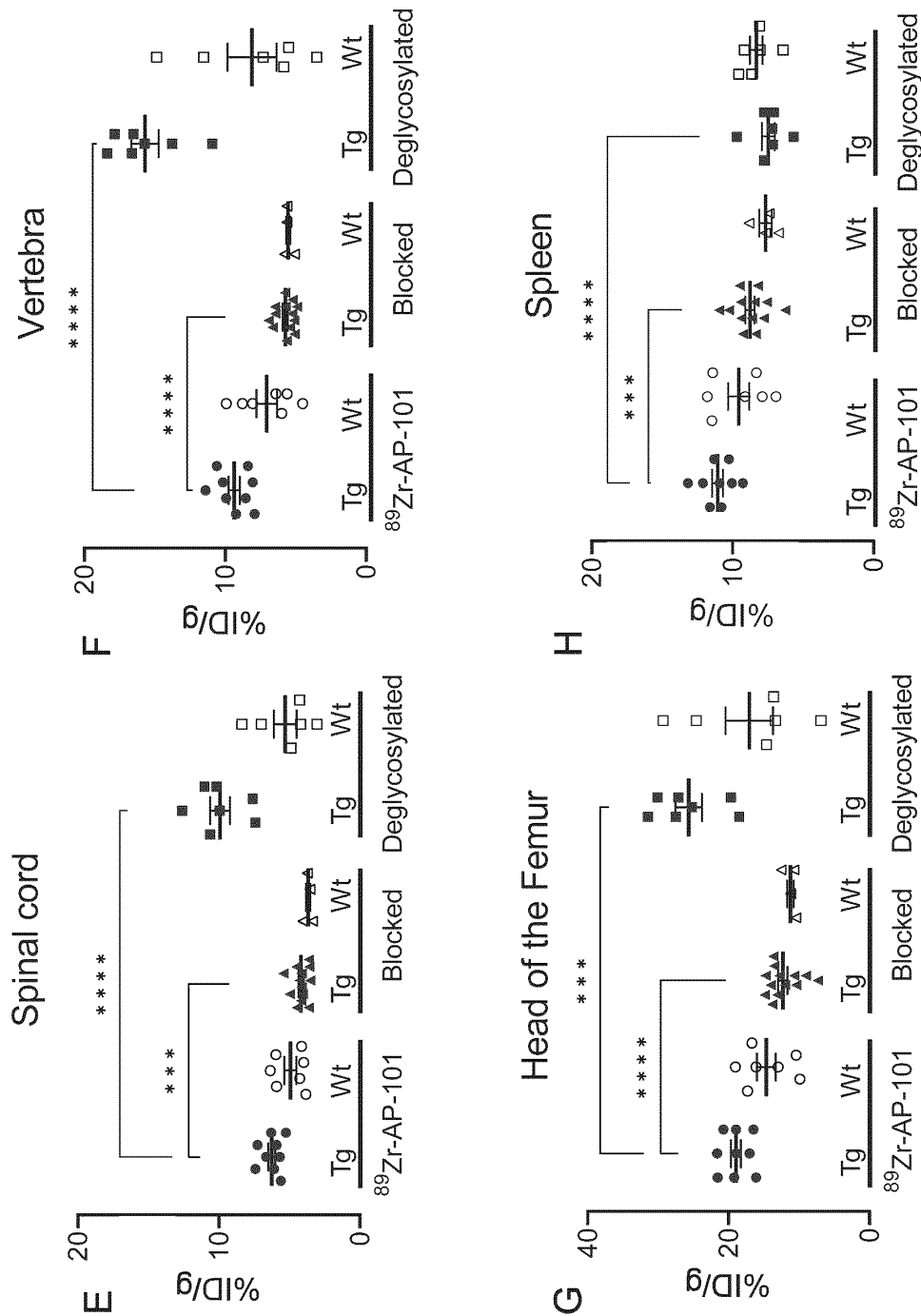


Fig. 3 (continued 2)

$\text{Corrected Activity} = \frac{\text{Measured activity} - \text{SF} \times \text{bone activity}}{\text{RC}}$	
Spot – L1	Mean activity (%ID/g) 8.677
Ventral bone ROI	10.471
Spill-in correction	6.583
Spill-in & RC correction	8.777

Thorn S.L. et al. J Nucl Med 2013; 54:1637–1644

Soret M. et al. J Nucl Med 2007; 48:932–945

Fig. 4

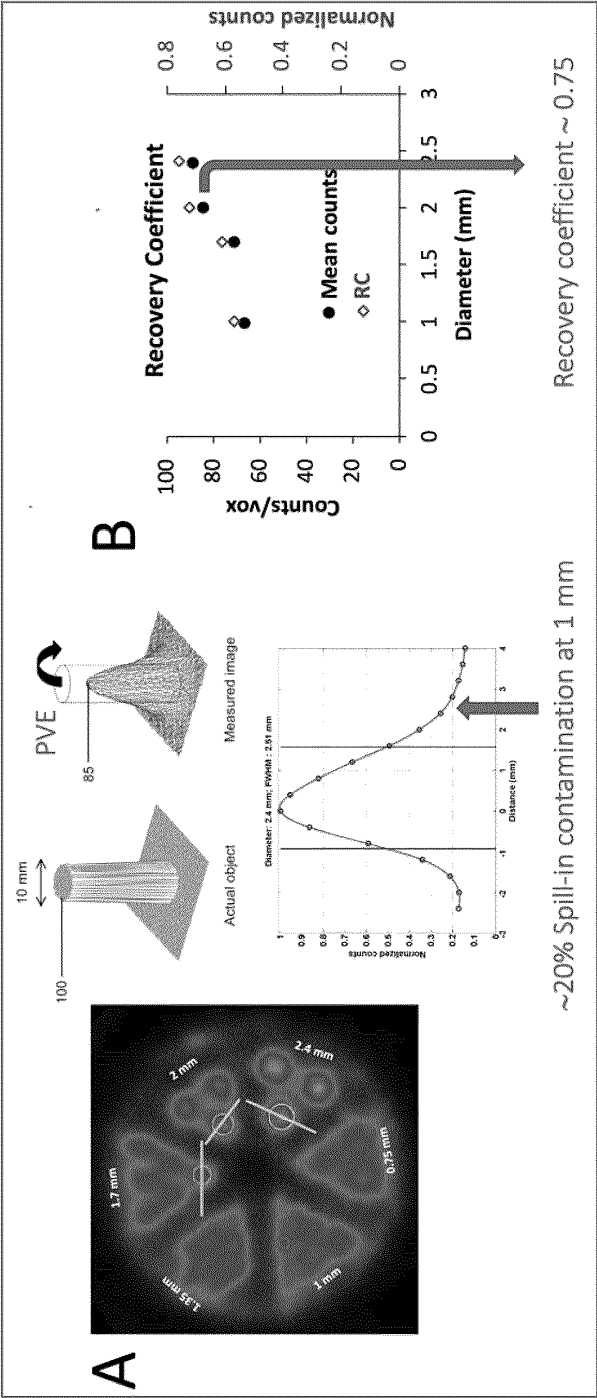


Fig. 5

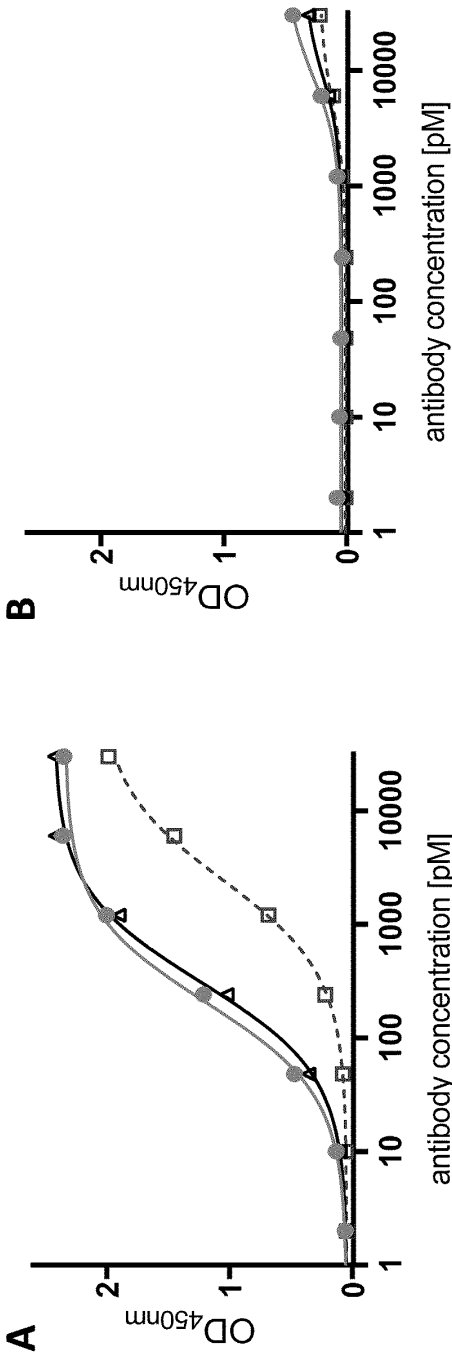


Fig. 6

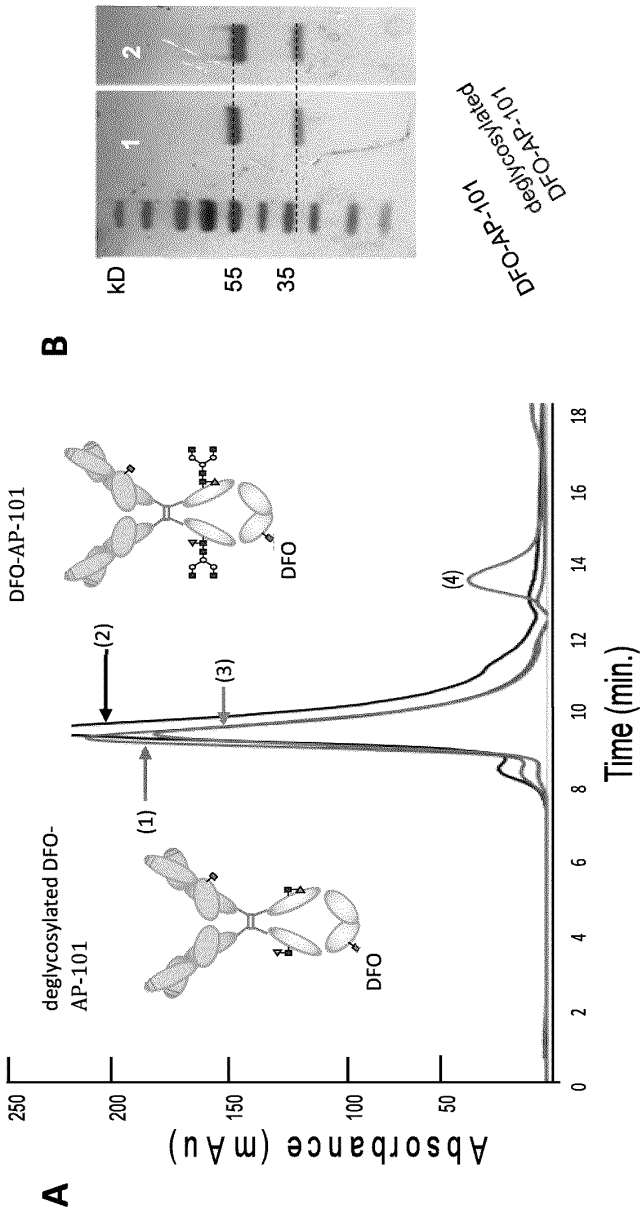


Fig. 7

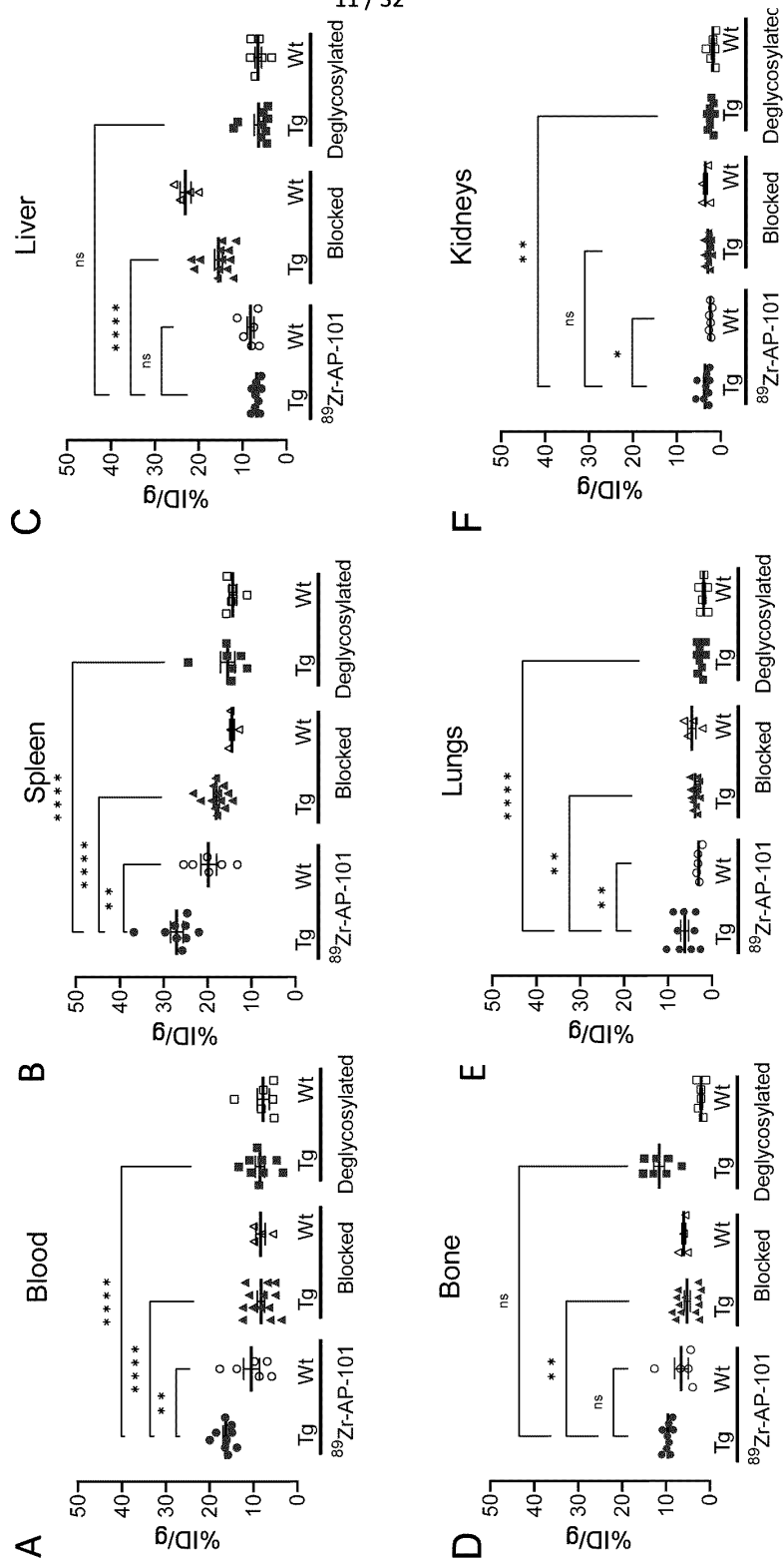


Fig. 8

1

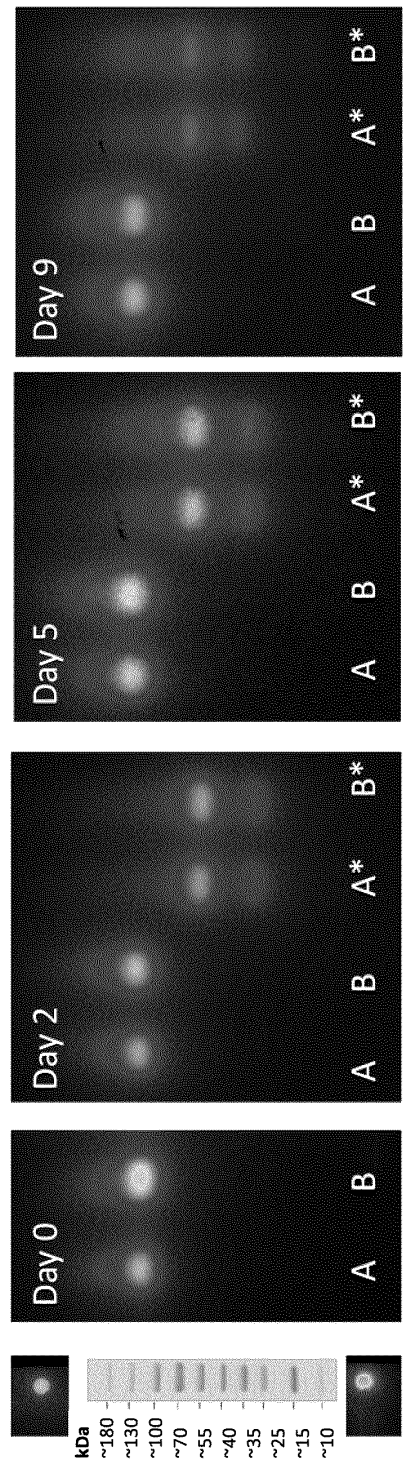


Fig. 9

Longitudinal study ⁸⁹ Zr-DFO-AP-101	Groups	Age	Conditions	Mice						
				Tg	ALS-001	ALS-002	ALS-003	ALS-004	ALS-005	
A	GROUP-1	91-101	⁸⁹ Zr-DFO-AP-101	Wt	CTL-001	CTL-002	CTL-003			G1
	GROUP-2	112-122	⁸⁹ Zr-DFO-AP-101	Tg	ALS-006	ALS-007	ALS-008	ALS-009	ALS-010	
				Wt	CTL-004	CTL-005	CTL-006			G2
	GROUP-3	126-136	⁸⁹ Zr-DFO-AP-101	Tg	ALS-011	ALS-012	ALS-013	ALS-014	ALS-015	G3
				Wt	CTL-007	CTL-008	CTL-009			
	GROUP-4	126-136		Tg	ALS-016	ALS-017	ALS-018	ALS-019	ALS-020	G4
				Wt	CTL-010	CTL-011	CTL-012	CTL-017		
	GROUP-5	126-136	Blocked	Tg	ALS-021	ALS-022	ALS-023	ALS-024	ALS-025	ALS-027
	GROUP-6	126-136		Tg	ALS-028	ALS-029	ALS-030	ALS-031	ALS-032	ALS-033
	GROUP-8	126-136		GROUP-8	Wt	CTL-013	CTL-014	CTL-015	CTL-016	G6
B	GROUP-7	126-136	Degly- ⁸⁹ Zr-DFO-AP-101	Tg	ALS-034	ALS-035	ALS-036	ALS-037	ALS-038	ALS-040
	GROUP-9	126-136		Wt	CTL-018	CTL-019	CTL-020	CTL-021	CTL-022	CTL-023
										G7
										ALS-041
										ALS-042
										ALS-043
										ALS-044
										ALS-045
										ALS-046
										ALS-047

Fig. 10

Descriptive statistics		Tg	Wt
Number of values		29	17
Minimum		23.10	27.20
25% Percentile		24.90	28.95
Median		26.30	30.10
75% Percentile		28.55	32.30
Maximum		33.40	35.40
Range		10.30	8.20
Mean		26.72	30.49
Std. Deviation		2.46	2.10
Std. Error of Mean		0.46	0.51
Coefficient of variation		9.2%	6.9%

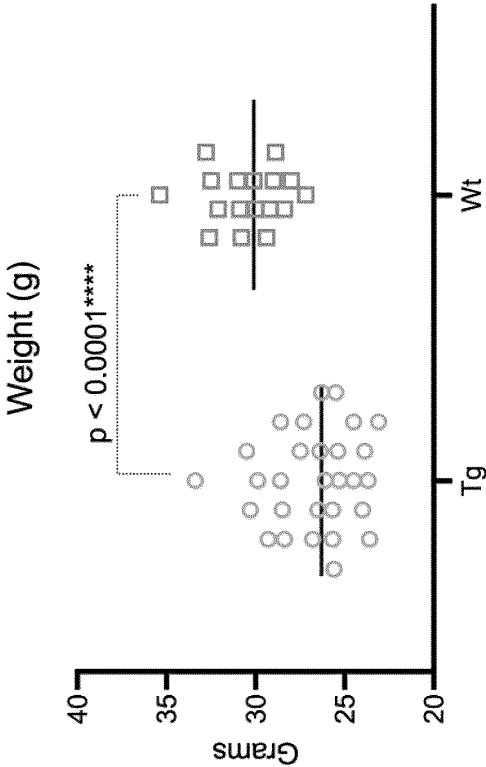
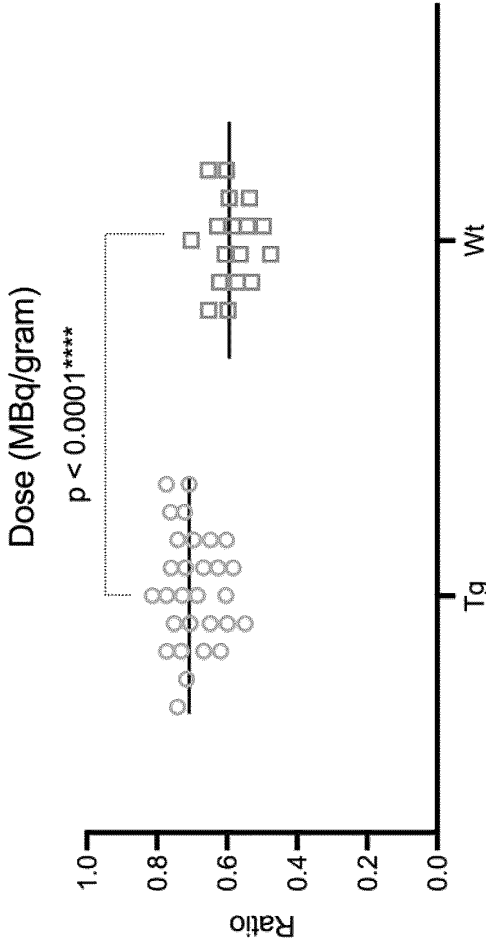


Fig. 11

Descriptive statistics		Tg	Wt
Number of values		29	17
Minimum		0.55	0.48
25% Percentile		0.64	0.54
Median		0.71	0.60
75% Percentile		0.75	0.62
Maximum		0.81	0.70
Range		0.27	0.23
Mean		0.69	0.59
Std. Deviation		0.07	0.06
Std. Error of Mean		0.01	0.01
Coefficient of variation		9.8%	10%



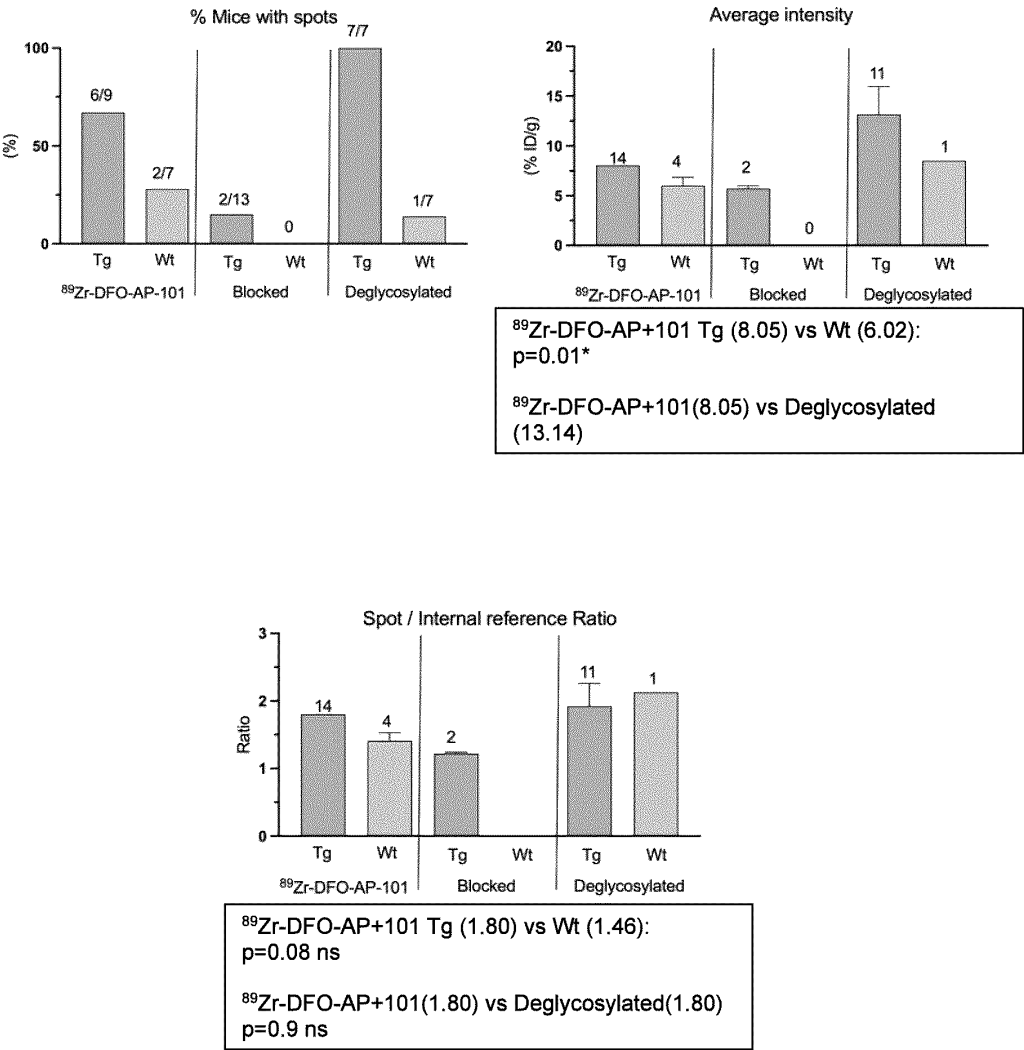


Fig. 14

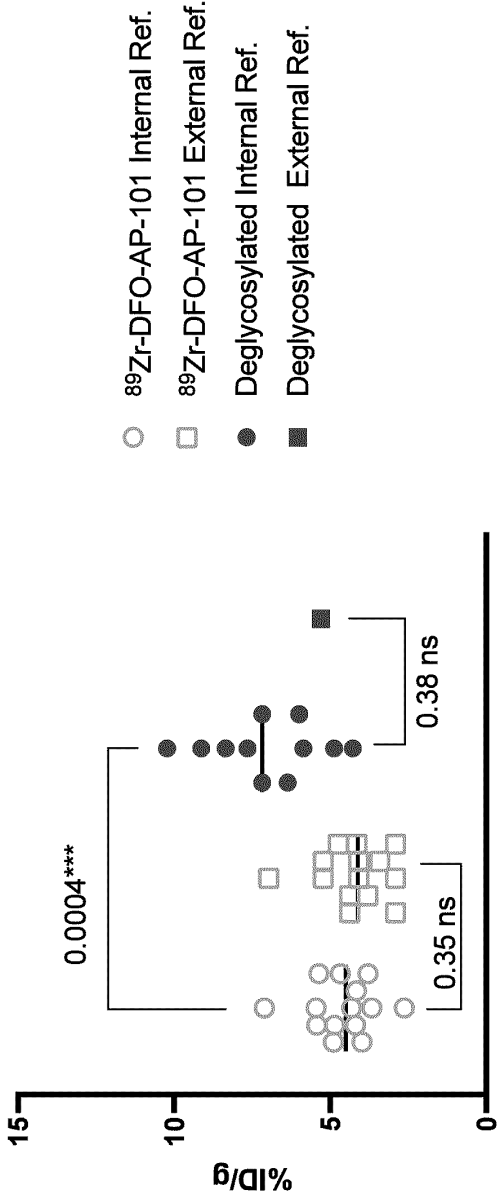


Fig. 15

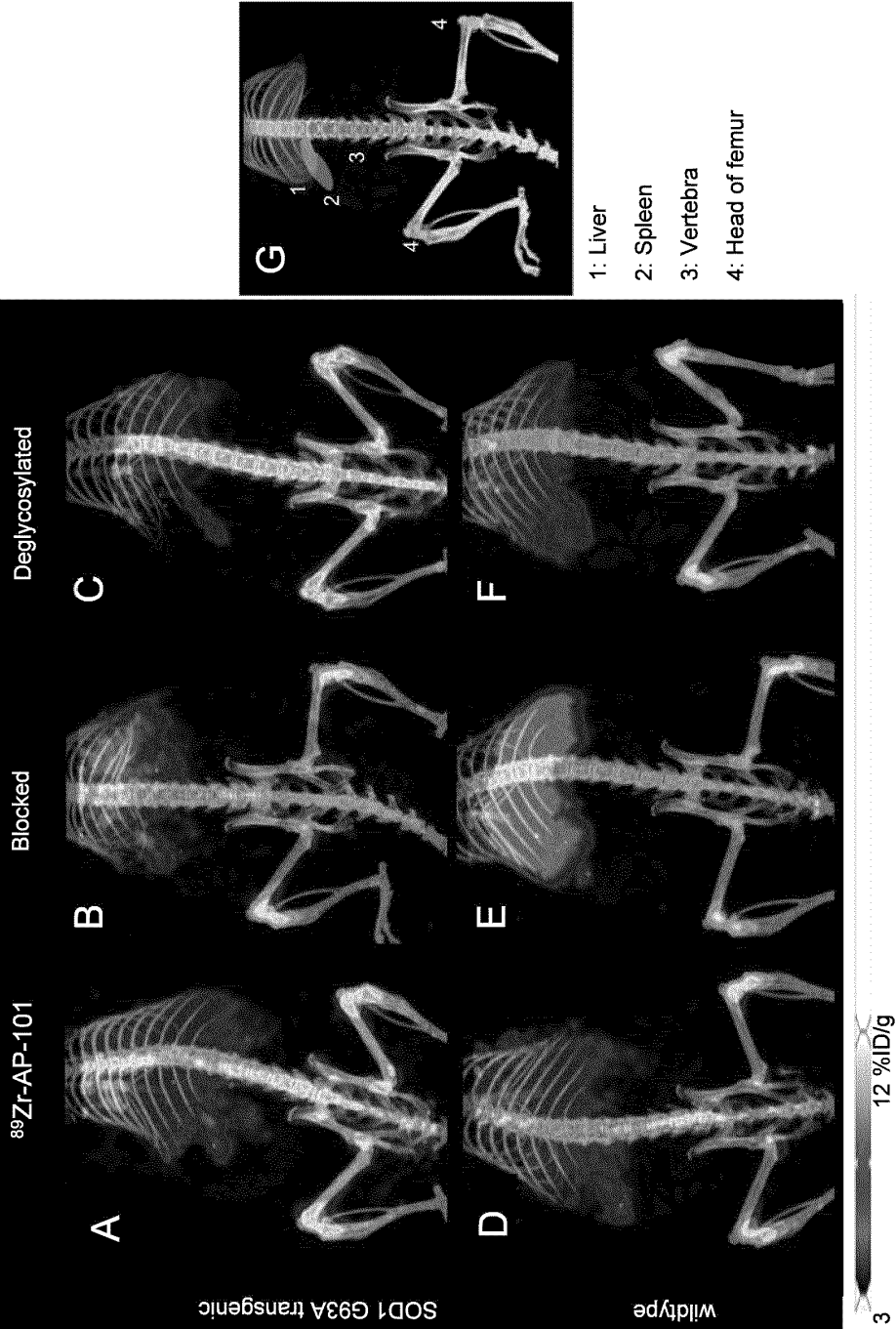


Fig. 16

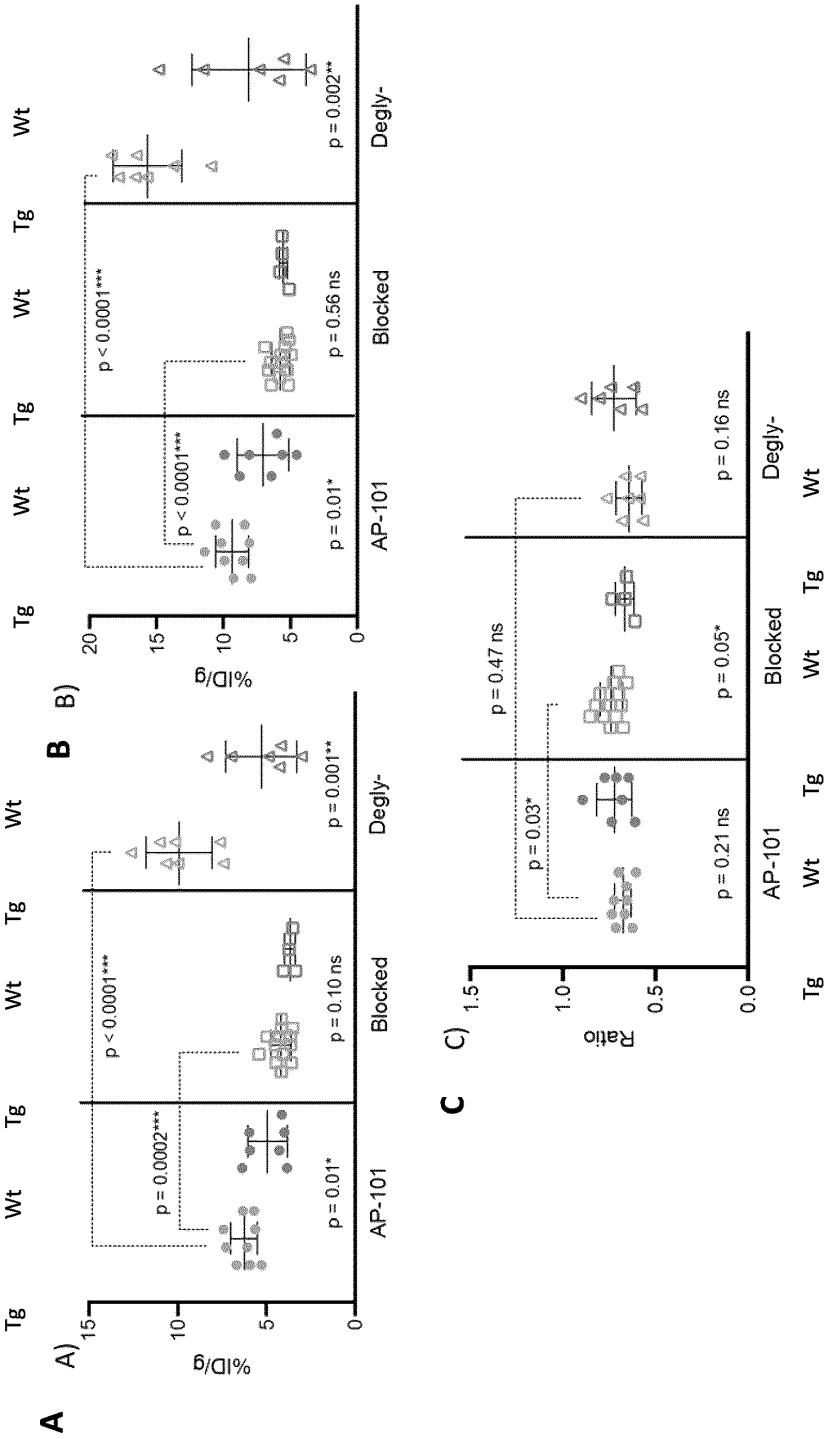


Fig. 17

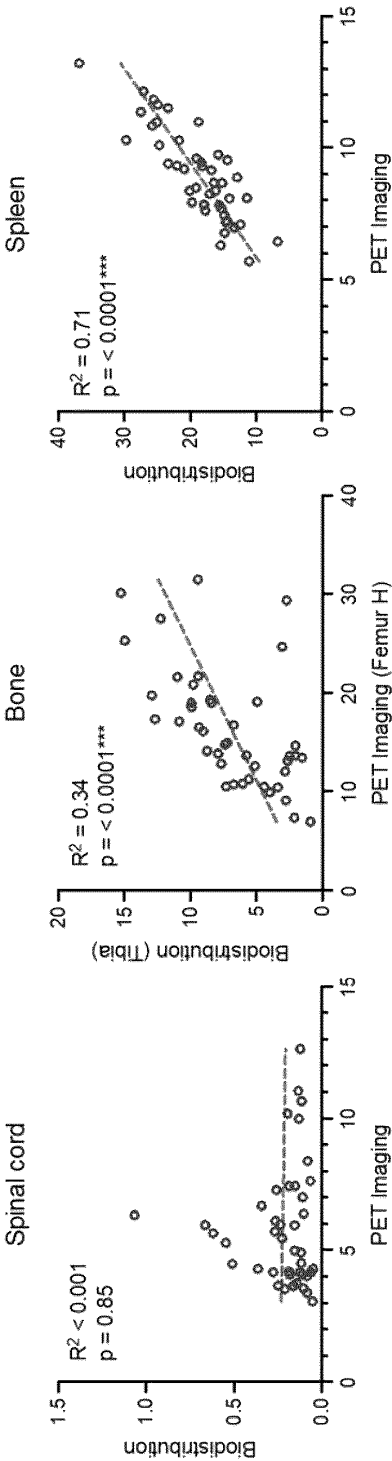
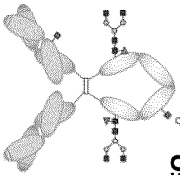
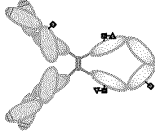


Fig. 18

	⁸⁹ Zr-DFO-AP101			Blocked (100 mg/kg) AP-101			Degly- ⁸⁹ Zr-DFO-AP101		
	Tg (ALS)	Wt (CTL)		Tg (ALS)	Wt (CTL)		Tg (ALS)	Wt (CTL)	
Number of mice	9	7		13	4		7	7	
Number of spots	14	4		2	0		11	1	
Number of mice with spots	6	2		2	0		7	1	
% of mice with spot	67% (6/9)	28% (2/7)		15% (2/13)	0%		100% (7/7)	14	
Spots/mouse	1.60 (14/9)	0.57 (4/7)		0.15 (2/13)	0		1.60 (11/7)	0.1	
Spots (Average %ID/g)	8.05 sdm 1.30	6.03 sdm 0.82		5.73 sdm 0.28	0		13.14 sdm 2.8	8.53	0.00
Internal Reference (Average %ID/g)	4.62 sdm 1.06	4.30 sdm 0.67		4.70 sdm 0.15	0		7.02 sdm 2.8	4.00	0.00
External Reference (Average %ID/g)	4.23 sdm 1.11	4.16 sdm 0.05		3.66	0		5.31	5.31	0.00
Spot/Internal reference (Ratio)	1.80 sdm 0.40	1.41 sdm 0.12		1.22 sdm 0.02	0		1.92 sdm 0.34	2.13	0.00
Spot/External Reference (Ratio)	2.01 sdm 0.58	1.45 sdm 0.19		1.56 sdm 0.08	0		2.47 sdm 0.53	1.61	0.00



DFO
⁸⁹Zr-DFO-AP-101



DFO
Degly-⁸⁹Zr-DFO-AP-101

Fig. 19

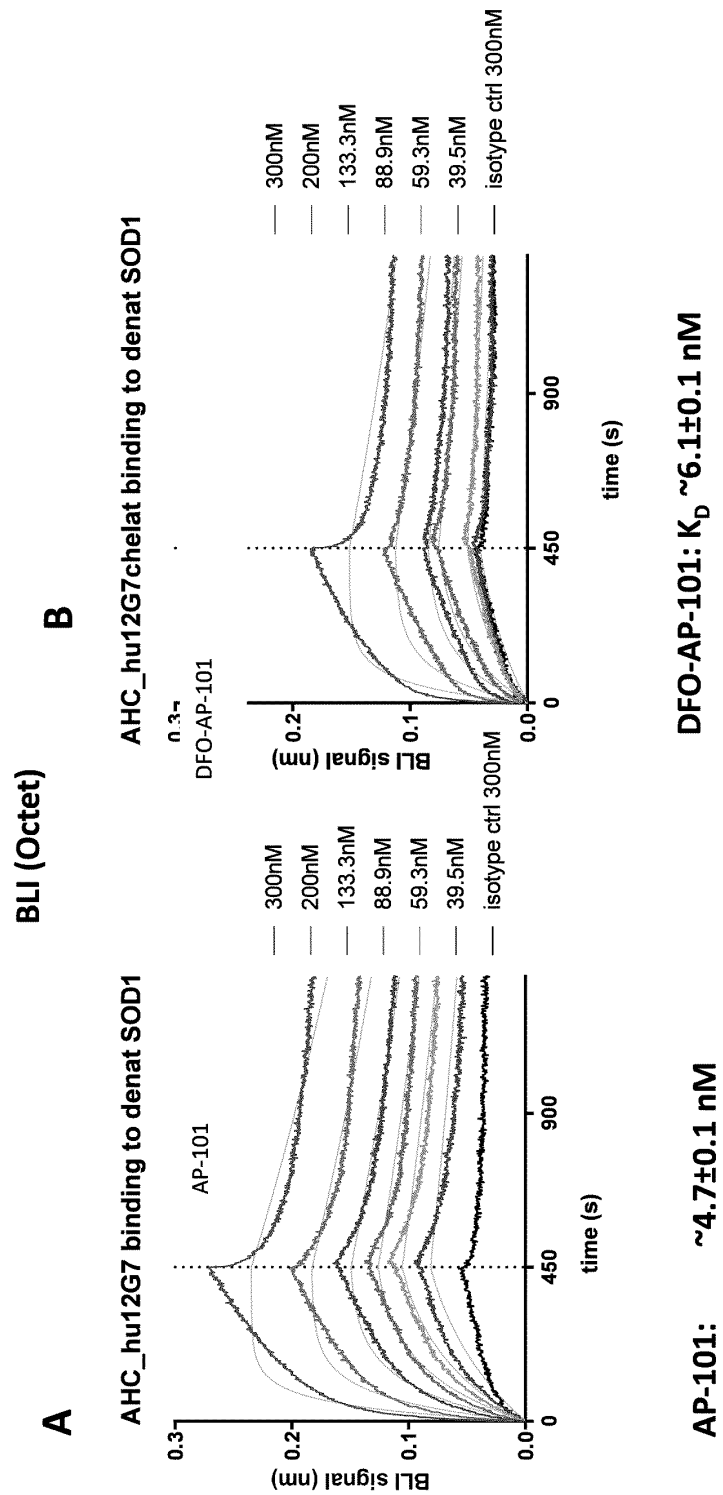


Fig. 20

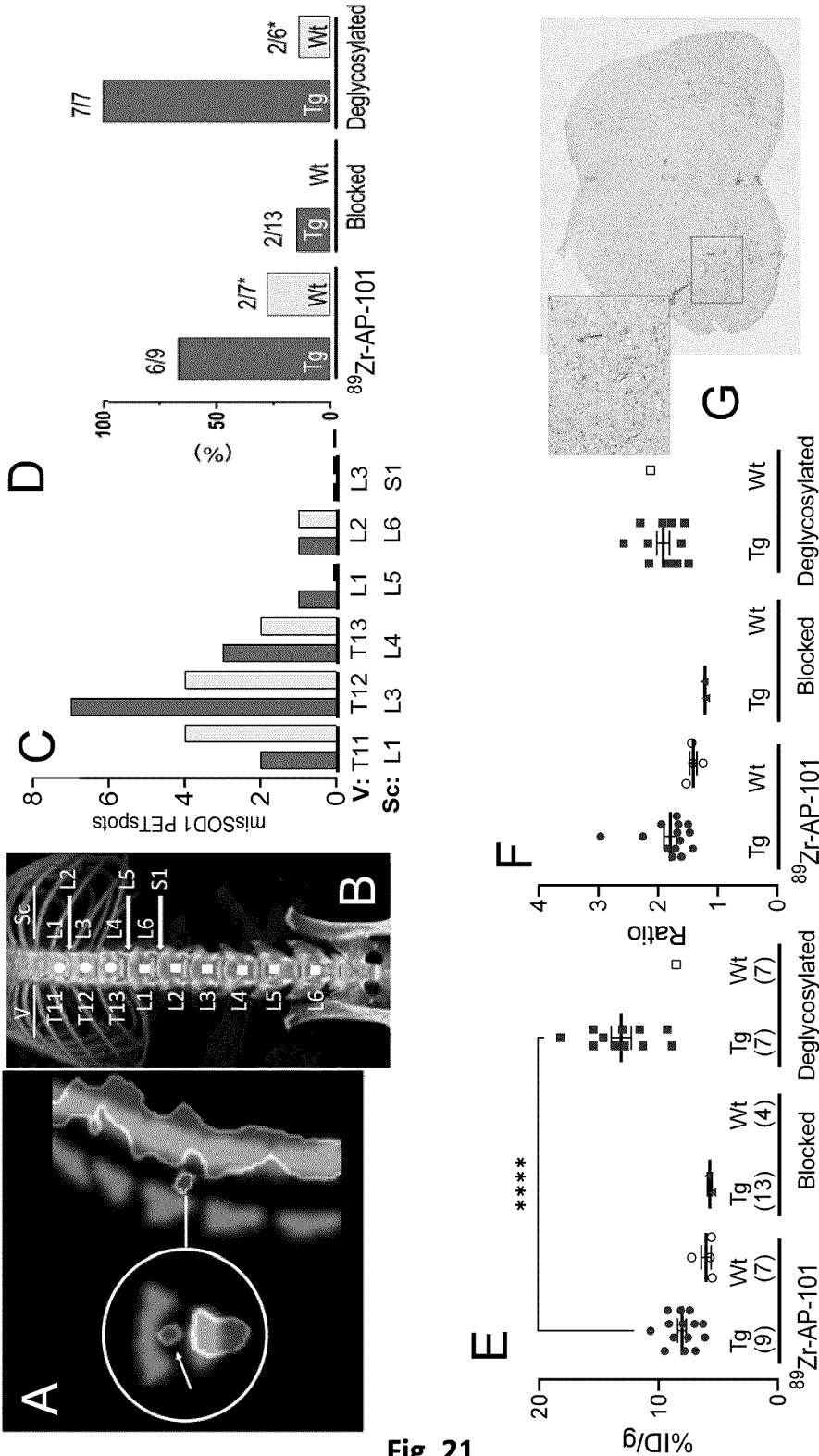


Fig. 21

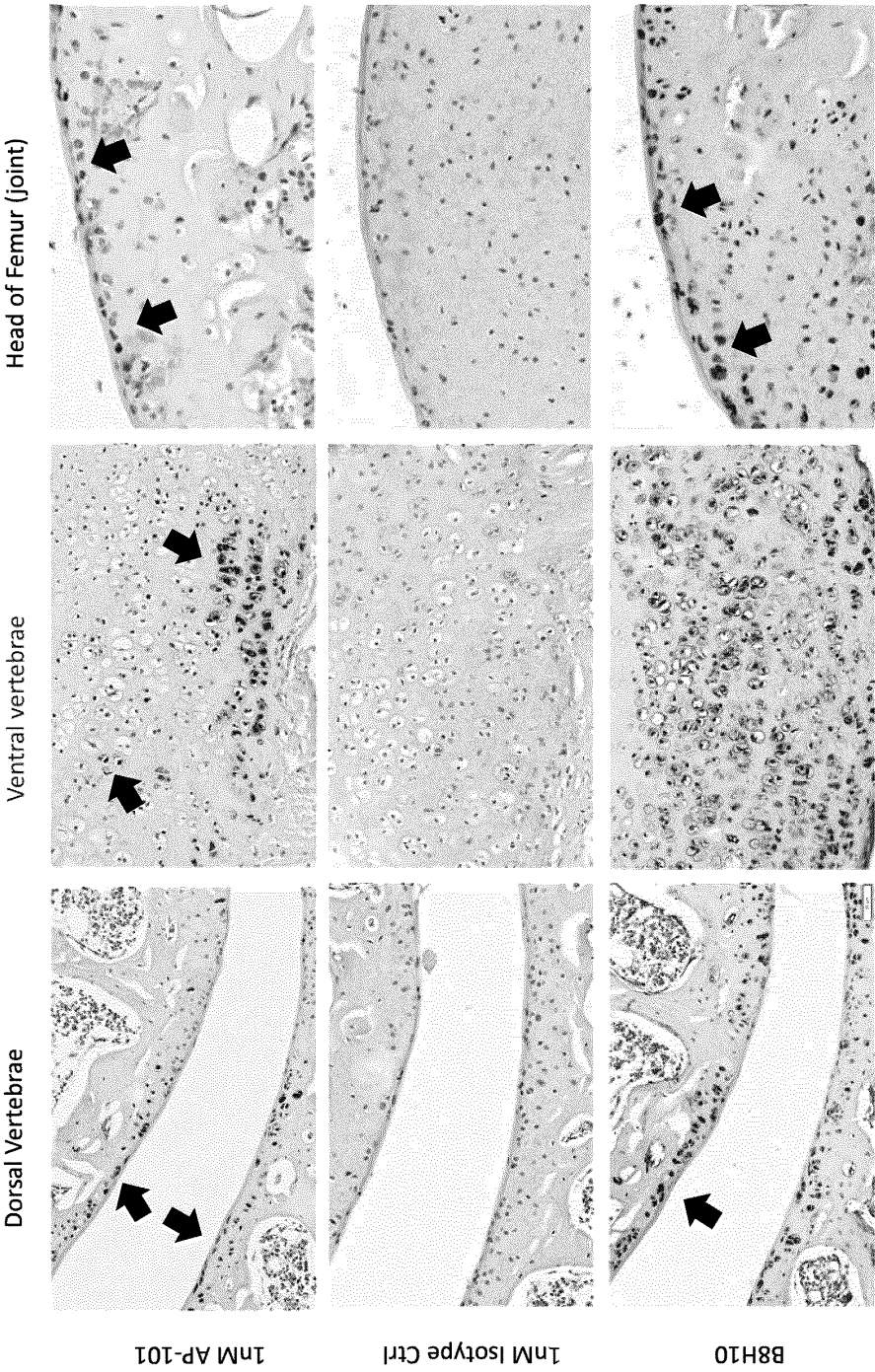


Fig. 22

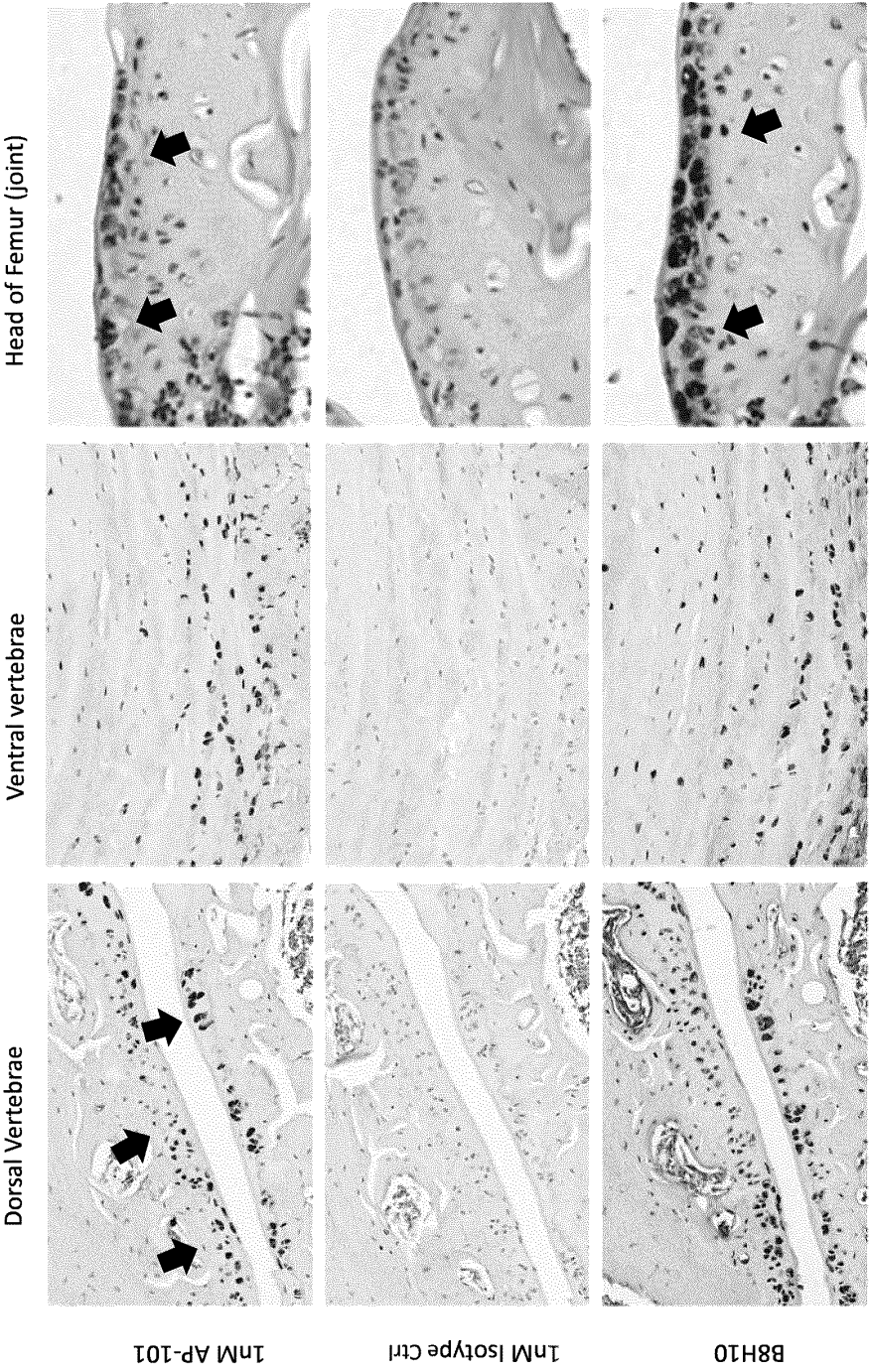


Fig. 22(continued)

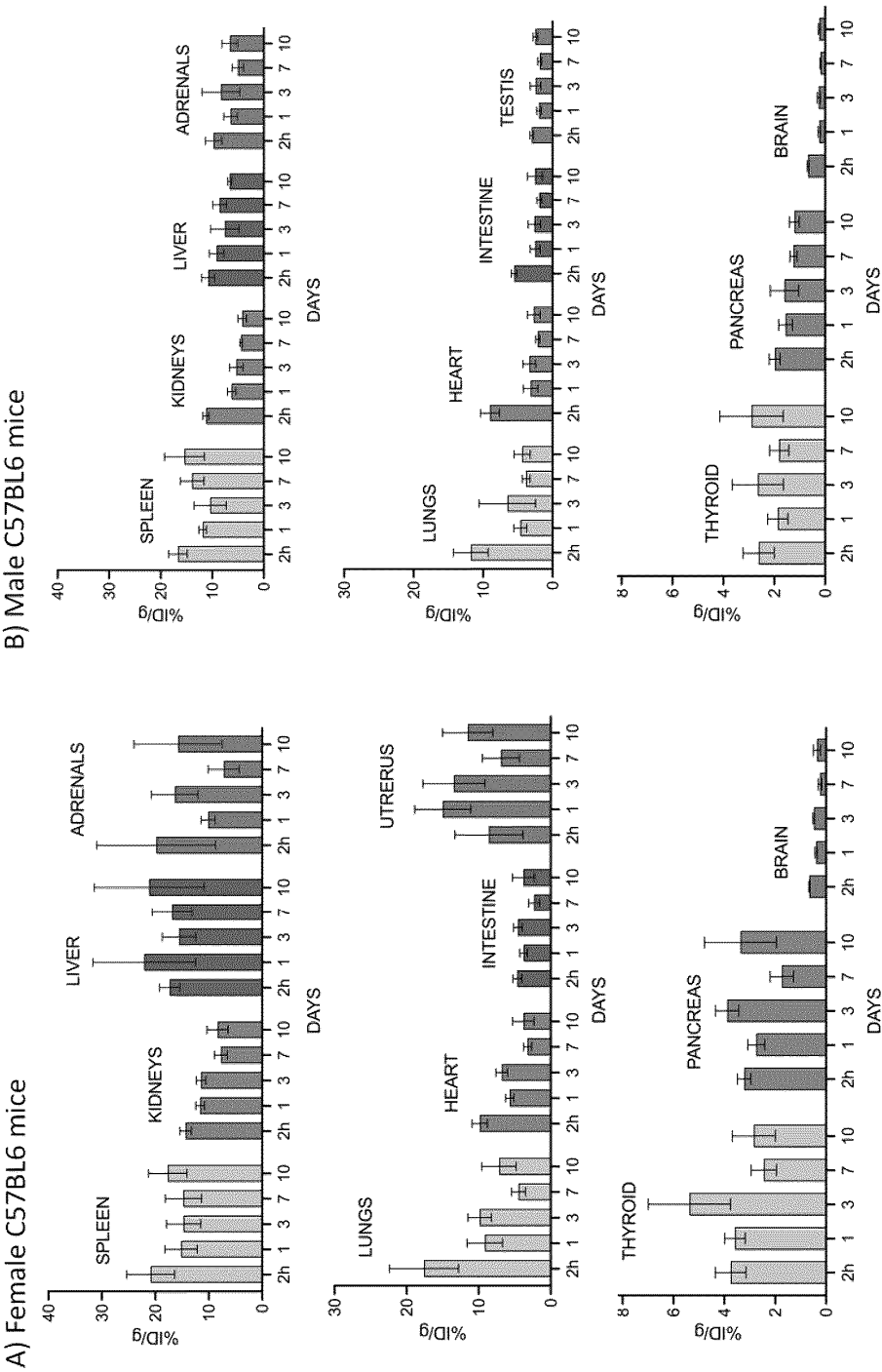


Fig. 23

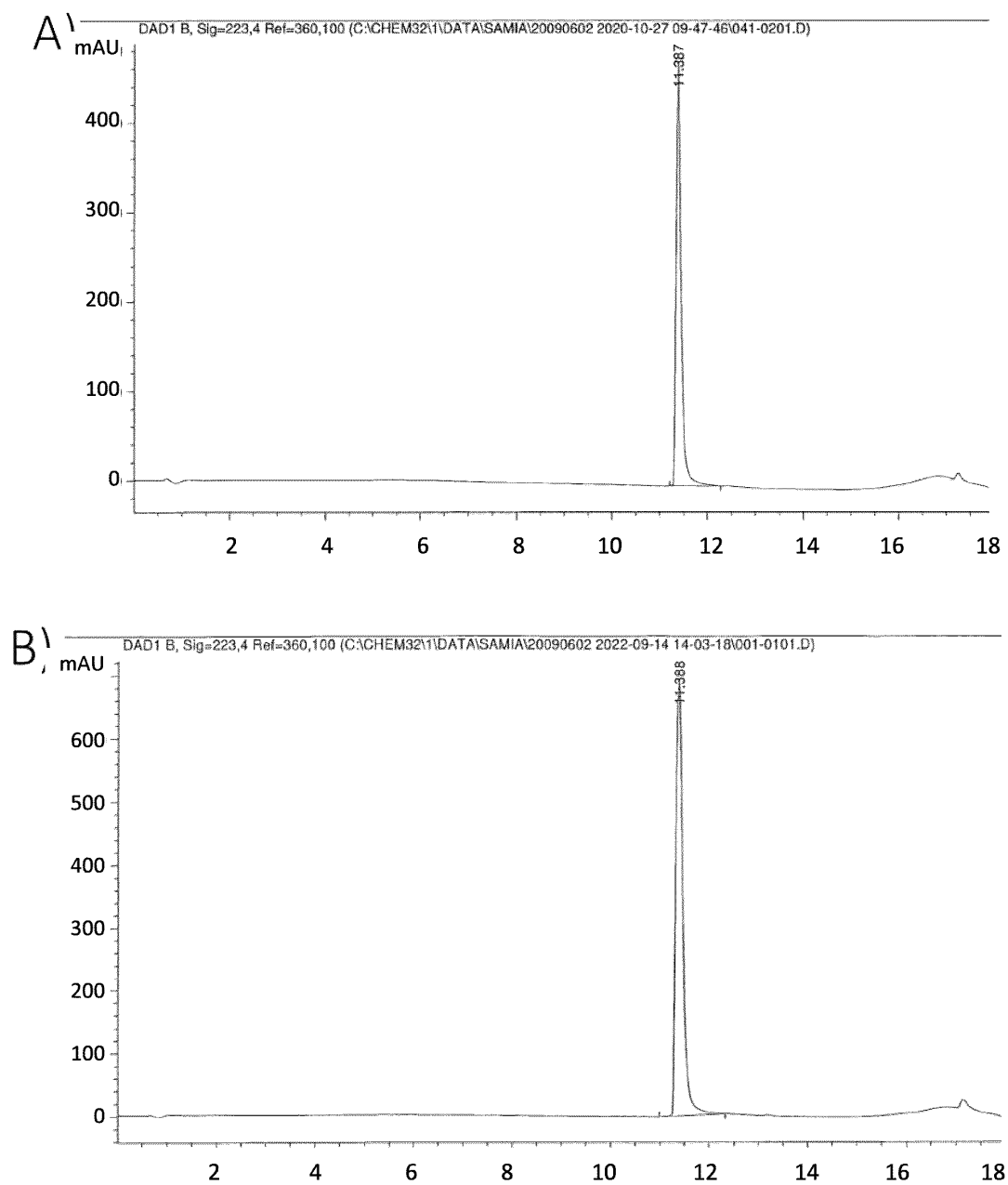


Fig. 24

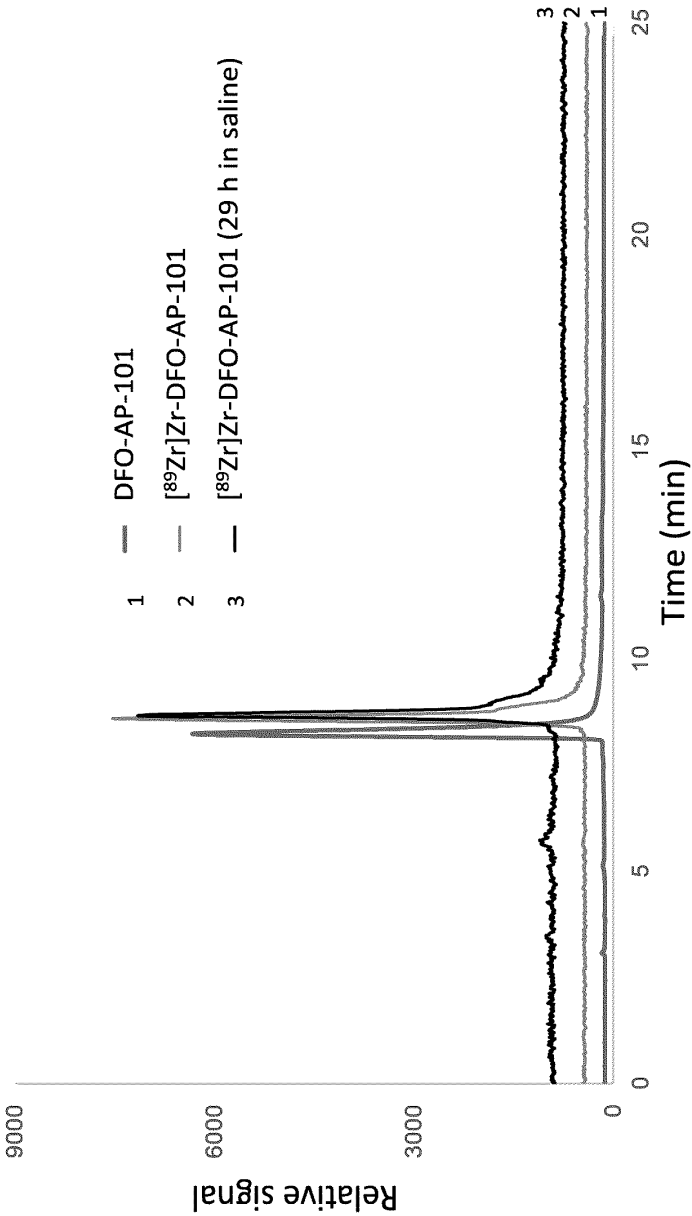


Fig. 25

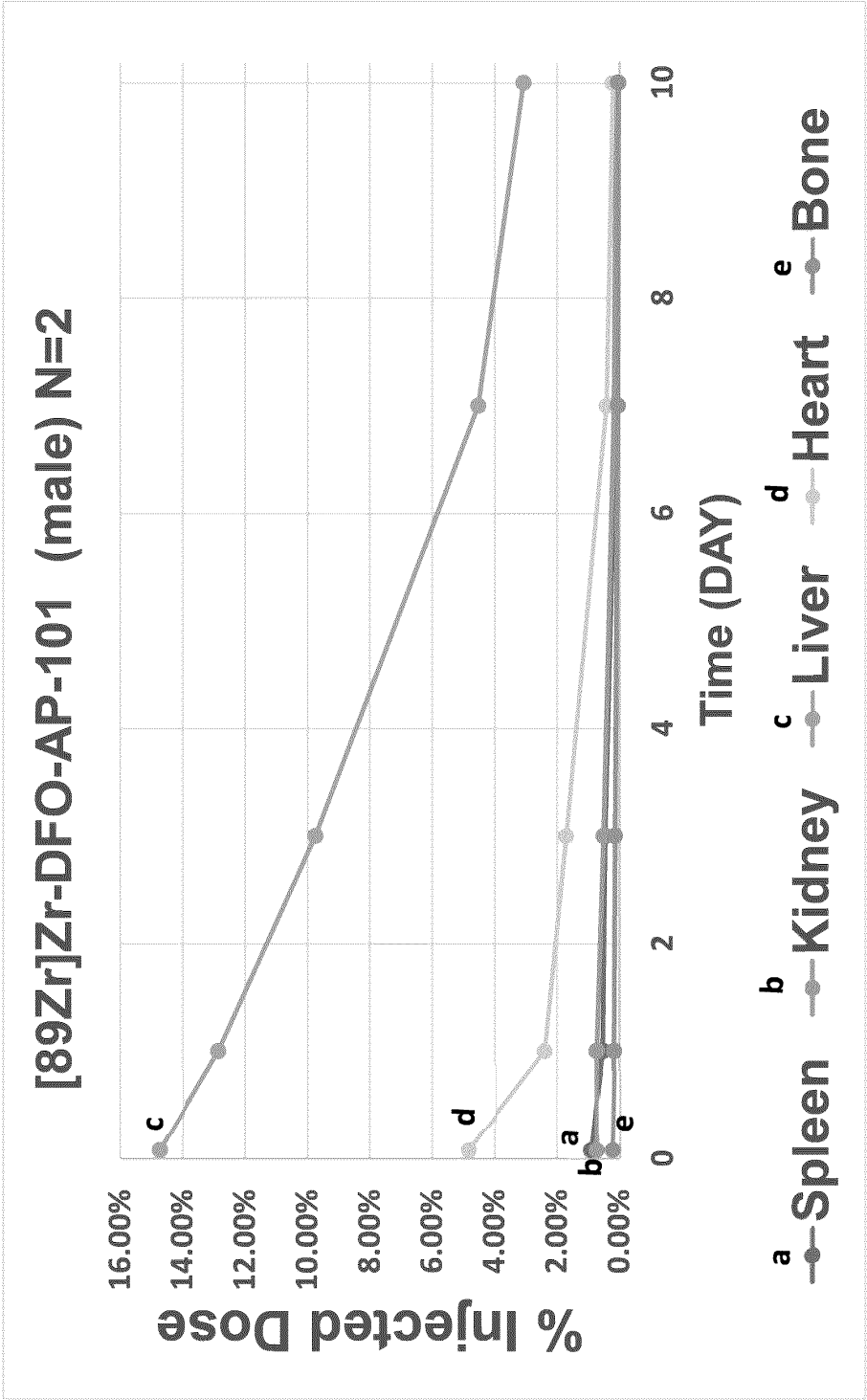


Fig. 26A

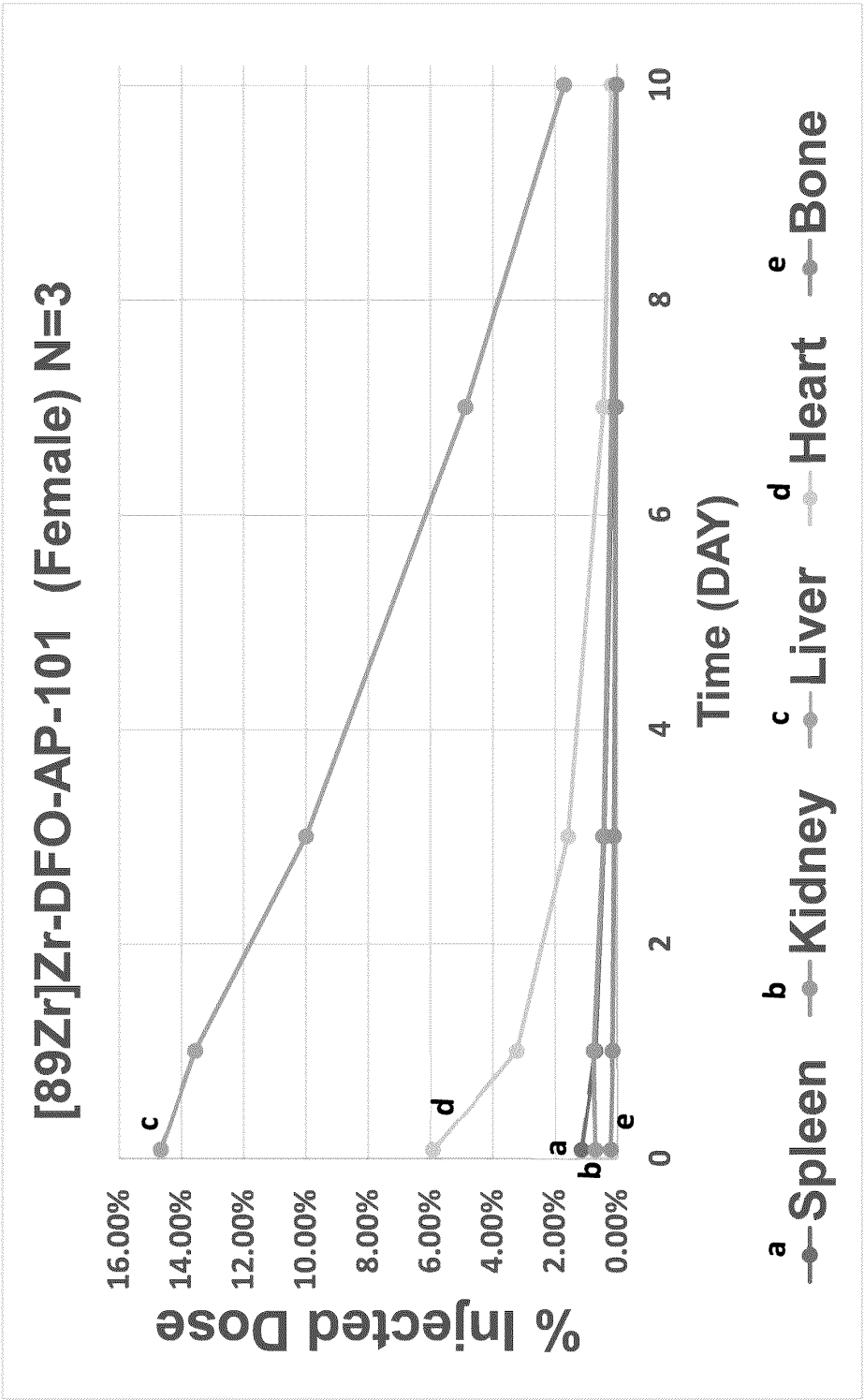


Fig. 26B

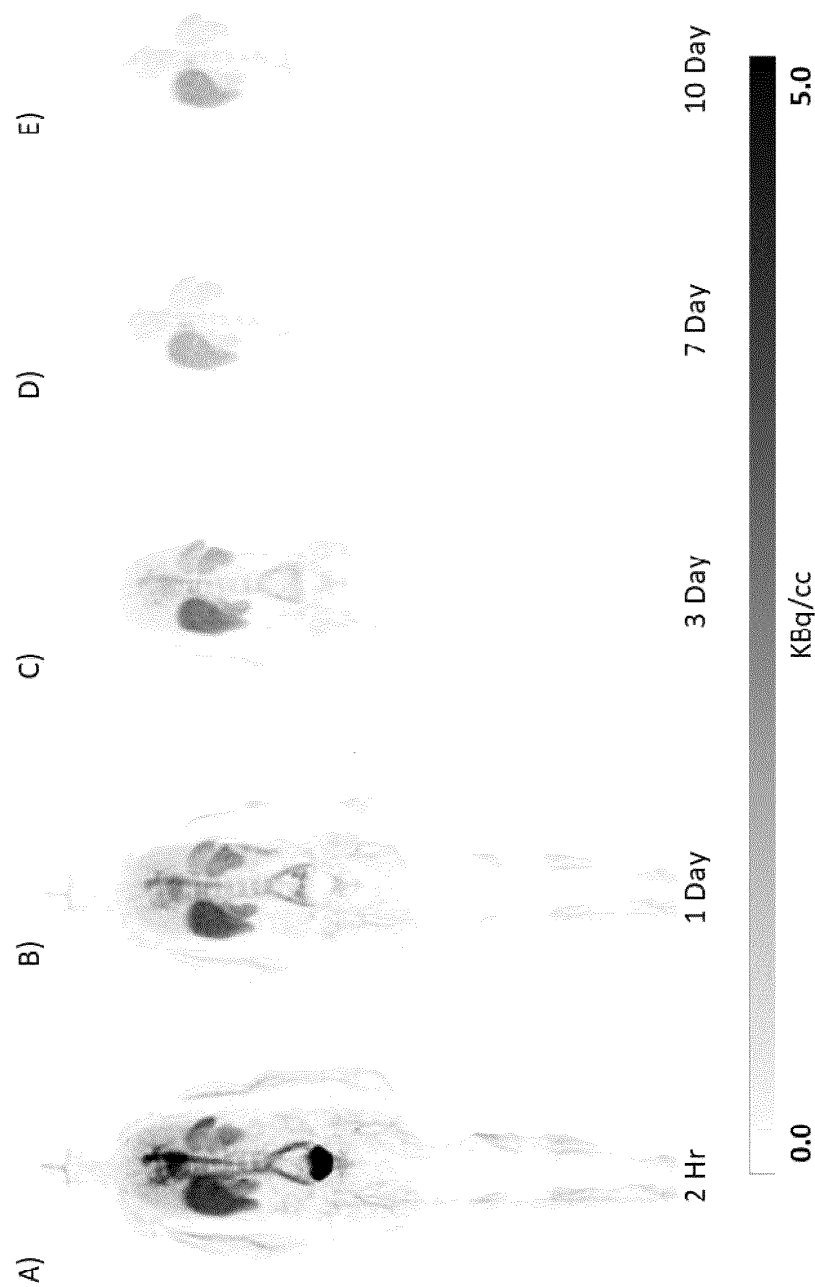


Fig. 27