

Appendix S1

Supplemental Table 1. Grade of severity for neurofibromatosis 1 by DNB Classification

Stage	Manifestations			Life function and social activity
	D	N	B	
1	1	0	0	No problems in daily life and social activity
2	1, 2	0 1	1 0, 1	Mild problems in daily life and social activity
3	3	0	0	Mild problems in daily life and severe problems in social activity
4	3	0 1	1 0, 1	Moderate problems in daily life and severe problems in social activity
5	4 any any	any 2 any	any any 2	Severe problems in daily life and social activity

Dermatological manifestations (D)

- 1 Pigmented macules and a few neurofibromas
- 2 Pigmented macules and many neurofibromas
- 3 Numerous neurofibromas (>1000, >1cm in size)
- 4 Large plexiform neurofibroma with severe secondary problems or malignant peripheral nerve sheath tumor

Neurological manifestations (N)

- 0 No neurological symptoms
- 1 Neurological symptoms (paralysis or pain) or/and abnormal neurological findings
- 2 Severe or progressive neurological symptoms (intellectual disability or brain tumor)

Bone manifestations (B)

- 0 No bone lesions
- 1 Mild or moderate bone lesions (deformity of spine or limbs) not requiring treatment
- 2 Severe bone lesions (dystrophic scoliosis, pseudoarthrosis or defect of skull or facial bone) requiring treatment