

Table S1. Characteristics of the 21 patients with keratitis-ichthyosis-deafness (KID) syndrome and fatal outcomes (20 patients reported in the literature and our case)

Patient number	1	2, 3, 4	5	6	7, 8	9	10	11	12	13	14	15	16	17	18	19	20	21	22
Reference	(2)	(2)	(2)	(2, 5)	(2)	(2)	(2)	(2)	(2)	(2)	(7)	(1)	(4)	(3)	(6)	(6)	(6)	(6)	(6)
Familial cases	-	2, dizygotic twins, 1 sibling	-	-	2, dizygotic twins	-	-	-	-	-	-	-	-	-	-	-	-	-	-
Origin	ND	African	Caucasian	Caucasian	African	ND	ND	Asian	ND	African	Caucasian	Japanese	South American	Japanese	ND	DN	DN	DN	DN
Suspected mode of inheritance	DN	Germ-line mosaic	DN	DN	Somatic mosaic	DN	DN	DN	DN	DN	DN	DN	DN	DN	DN	DN	DN	DN	DN
Age at onset	B	B	2 M	B	B	B	B	B	B	1M	B	B	B	B	B	B	B	B	B
Dystrophic nails	+	+	-	+	+	-	+	+	+	+	+	+	+	-	ND	ND	ND	ND	ND
Hyperkeratosis	+	+	+	+	+	ND	-	+	+	+	+	-	+	+	+	+	+	+	+
Keratitis	-	-	+	+	-	-	+	+	+	+	+	+	ND	+	+	+	+	+	+
Hearing deficit	+	ND	+	+	ND	ND	+	+	+	ND	+	ND	ND	+	+	+	+	+	+
Other anomalies	+	+	-	-	+	+	-	-	-	-	-	-	+	+	+	+	+	+	+
Age at death	3 M	1 M, 1 M, 5 M	12 M	6 M	30 D, 5 M	2 M	3 M	51 D	77 D	16 M	29 Y	ND (<18M)	9 D	33 Y	1.75 M	6 M	2M	3W	7 Y
Cause of death	S	S	S	S	S	S	S	S/P	S	S	S	ND	S	P	S	S	O	ND	S
Antibiotic prophylaxis	-	+	-	+	-	-	-	-	-	-	-	ND	-	-	ND	ND	ND	ND	ND
GJB2 mutation	p.Ala88Val	p.Gly45Glu	p.Gly45Glu	p.Gly45Glu	p.Gly45Glu	p.Gly45Glu	p.Ser17Phe	p.Ser17Phe	ND	ND	p.Ser17Phe	p.Gly45Glu	p.Ala88Val	p.Asp50Asn	p.Ala88Val	p.Ala88Val	p.Ala88Val	p.Ala88Val	p.Ala88Val

*Congenital abnormality of the brainstem.

B: birth; D: day; DN: *de novo*; M: month; ND: not determined; O: other*; P: pneumonia; S: septicemia; SCC: spinous cell carcinoma; W: week; Y: year.