

Table S2

Figure 2B	IF_Collagen	$F_{ARA} = 18.06$ $P < 0.001^{***}$				
			Mean (sd)	n	Significance	p-value
	Control	rHSP47+ vs. rHSP47-	730.74(578.03) vs. 114.34(107.98)	n=8, n=8	*	$P=0.0460$
	Proband 1	rHSP47+ vs. rHSP47-	319.96(191.24) vs. 1000.05(550.94)	n=8, n=8	**	$P=0.0063$
	Proband 2	rHSP47+ vs. rHSP47-	242.19(148.23) vs. 2104.76(1136.72)	n=8, n=8	***	$P < 0.001$
	Proband 3	rHSP47+ vs. rHSP47-	2104.76(1136.72) vs. 1878.13(777.98)	n=8, n=8	***	$P < 0.001$
	Control vs. Proband 1	rHSP47-	11.13(8.76) vs. 42.63(9.71)	n=8	***	$P < 0.001$
	Control vs. Proband 2	rHSP47-	11.13(8.76) vs. 54.13(8.66)	n=8	***	$P < 0.001$
	Control vs. Proband 3	rHSP47-	11.13(8.76) vs. 54.00(6.97)	n=8	***	$P < 0.001$

Figure 2C	IF_PDI	$F_{ARA} = 15.12$ $P < 0.001^{***}$				
	Control	rHSP47+ vs. rHSP47-	784.08(511.28) vs. 387.58(490.98)	n=8, n=8	*	$P=0.0357$
	Proband 1	rHSP47+ vs. rHSP47-	314.57(148.22) vs. 1092.04(304.16)	n=8, n=8	***	$P=0.001$
	Proband 2	rHSP47+ vs. rHSP47-	1144.81(534.40) vs. 1825.78(512.80)	n=8, n=8	*	$P=0.0274$
	Proband 3	rHSP47+ vs. rHSP47-	318.00(180.58) vs. 1527.63(520.18)	n=8, n=8	***	$P < 0.001$
	Control vs. Proband 1	rHSP47-	14.38(18.09) vs. 39.88(8.37)	n=8	***	$P < 0.001$
	Control vs. Proband 2	rHSP47-	14.38(18.09) vs. 54.88(8.68)	n=8	***	$P < 0.001$
	Control vs. Proband 3	rHSP47-	14.38(18.09) vs. 49.00(11.44)	n=8	***	$P < 0.001$

Figure 2D	IF_PDI_Collagen	$F_{ARA} = 40.35$ $P < 0.001^{***}$				
	Control	rHSP47+ vs. rHSP47-	111.7(130.25) vs. 19.53(20.99)	n=8, n=8	*	$P=0.0272$

	Proband 1	rHSP47+ vs. rHSP47-	267.11(190.68) vs. 1006.60(350.61)	n=8, n=8	**	P=0.0016
	Proband 2	rHSP47+ vs. rHSP47-	350.90(289.35) vs. 1689.75(822.62)	n=8, n=8	***	P=0.001
	Proband 3	rHSP47+ vs. rHSP47-	143.88(45.03) vs. 1937(605.29)	n=8, n=8	***	P<0.001
	Control vs. Proband 1	rHSP47-	6.5(4.42) vs. 45.88(5.86)	n=8, n=8	***	P<0.001
	Control vs. Proband 2	rHSP47-	6.5(4.42) vs. 54.00(6.79)	n=8, n=8	***	P<0.001
	Control vs. Proband 3	rHSP47-	6.5(4.42) vs. 56.63(6.19)	n=8, n=8	***	P<0.001

Figure 3B	Thioflavin	$F_{ARA}= 86.82$ $P<0.001^{***}$					
		Control	rHSP47+ vs. rHSP47-	1.33 (1.07) vs. 3.34 (2.55)	n=7, n=7	Ns.	P=0.1417
		Proband 1	rHSP47+ vs. rHSP47-	183.15(121.15) vs. 922.3526(169.248)	n=7, n=7	**	P=0.0017
		Proband 2	rHSP47+ vs. rHSP47-	11.36(11.30) vs. 592.03(160.87)	n=7, n=7	**	P=0.0017
		Proband 3	rHSP47+ vs. rHSP47-	89.36(103.18) vs. 446.90(109.14)	n=7, n=7	**	P=0.0027
		Control vs. Proband 1	rHSP47-	10.71(5.91) vs. 52.43(2.88)	n=7	***	P<0.001
		Control vs. Proband 2	rHSP47-	10.71(5.91) vs. 45.14(5.08)	n=7	***	P<0.001
		Control vs. Proband 3	rHSP47-	10.71(5.91) vs. 39.86(3.98)	n=7	***	P<0.001
Figure 3D	ER thickness	Proband 1	rHSP47+ vs. rHSP47-	135.99(42.87) vs. 347.85(114.00)	n=30, n=30	***	MW p<0.001
		Proband 2	rHSP47+ vs. rHSP47-	155 (67) vs. 302 (92)	n=30, n=31	***	MW p<0.001
		Proband 3	rHSP47+ vs. rHSP47-	131 (58) vs. 270 (68)	n=30, n=30	***	MW p<0.001
Figure 4B	Cell (%)						
	Live	Control	rHSP47+ vs. rHSP47-	79.14(1.41) vs. 84.83(2.60)	n=3, n=3	*	MW P<0.04
		Proband 1	rHSP47+ vs.	66.74(1.21) vs.	n=3, n=3	*	MW P<0.04

			rHSP47-	57.56(1.7)8			
		Proband 2	rHSP47+ vs. rHSP47-	79.14 (1.41) Vs. 84.83 (2.60)	n=3, n=3	Ns.	MW P=0.1
		Proband 3	rHSP47+ vs. rHSP47-	86.72 (0.63) Vs. 87.30 (0.60)	n=3, n=3	Ns.	MW P=0.4
	Live	Control vs. Proband 1	rHSP47-	84.83(2.60) vs. 57.56(1.78)	n=3, n=3	*	P=0.04
		Control vs. Proband 2	rHSP47-	84.83(2.60) Vs. 87.30(0.60)	n=3, n=3	Ns.	P=0.2
		Control vs. Proband 3	rHSP47-	84.83(2.60) Vs. 86.86(0.45)	n=3, n=3	Ns.	P=0.7
	Apoptotic	Control	rHSP47+ vs. rHSP47-	11.35(0.83) vs. 10.35(2.00)	n=3, n=3	Ns.	MW P=0.7
		Proband 1	rHSP47+ vs. rHSP47-	20.97(1.12) vs 31.29(0.99)	n=3, n=3	*	MW P<0.01
		Proband 2	rHSP47+ vs. rHSP47-	9.61 (0.33) Vs 9.15 (0.28)	n=3, n=3	Ns.	MW P=0.2
		Proband 3	rHSP47+ vs. rHSP47-	10.72 (0.81) vs. 11.56 (0.41)	n=3, n=3	Ns.	MW P=0.4
	Apoptotic	Control vs. Proband 1	rHSP47-	10.35(2.00) vs. 31.29(0.99)	n=3, n=3	**	MW P=0.004
		Control vs. Proband 2	rHSP47-	10.35 (2.00) vs 9.15 (0.29)	n=3, n=3	Ns.	MW P=0.71
		Control vs. Proband 3	rHSP47-	10.35 (2.00) vs. 11.56 (0.41)	n=3, n=3	Ns.	MW P=0.7
Figure 4D	Live	Control	rHSP47+ vs. rHSP47-	89.67(2.90) vs. 72.75(0.73)	n=3, n=3	*	MW P<0.049
		Proband	rHSP47+ vs. rHSP47-	16.80(1.54) vs. 8.53(0.45)	n=3, n=3	*	MW P<0.04
	Live	Control vs. Proband	rHSP47-	72.75(0.73) vs. 8.53(0.45)	n=3, n=3	**	MW P=0.04
	Early apoptotic	Control	rHSP47+ vs. rHSP47-	8.65(0.14) vs. 19.78(1.05)	n=3, n=3	*	MW P<0.049
		Proband	rHSP47+ vs. rHSP47-	67.18(3.75) vs. 25.16(2.87)	n=3, n=3	*	MW P<0.049
	Early apoptotic	Control vs. Proband	rHSP47-	8.65(0.14) vs. 19.78(1.05)	n=3, n=3	*	MW P=0.049
	Late apoptotic	Control	rHSP47+ vs. rHSP47-	2.72(0.064) vs. 5.43(0.317)	n=3, n=3	*	MW P<0.0463
		Proband	rHSP47+ vs. rHSP47-	15.83(2.98) vs. 66.45(2.19)	n=3, n=3	*	MW P<0.049
	Late apoptotic	Control vs. Proband	rHSP47-	5.43(0.32) vs. 66.45(2.18)	n=3, n=3	*	MW P=0.046

Figure 5A	Secreted collagen	Control	rHSP47+ vs. rHSP47-	158.72(13.83) vs. 106.37(16.27)	n=3, n=3	*	MW P=0.049
		Proband 1	rHSP47+ vs. rHSP47-	56.82(12.89) vs. 34.04(1.32)	n=3, n=3	*	MW P=0.049
		Proband 2	rHSP47+ vs. rHSP47-	158.24(24.80) vs. 84.75(7.63)	n=3, n=3	*	MW P=0.049
		Proband 3	rHSP47+ vs. rHSP47-	76.83(7.38) vs. 49.70(3.98)	n=3, n=3	*	MW P=0.049
		Control vs. Proband 1	rHSP47-	106.37(16.28) vs. 34.04(1.32)	n=3, n=3	***	P<0.001
		Control vs. Proband 2	rHSP47-	106.37(16.28) vs. 84.75(7.63)	n=3, n=3	*	P=0.049
		Control vs. Proband 3	rHSP47-	106.37(16.28) vs. 49.70(3.98)	n=3, n=3	***	P<0.001
		KW=17.92 p<0.001					
Figure 5F	Matrix collagen	Control	rHSP47+ vs. rHSP47-	129.32(19.85) vs. 123.65(22.95)	n=3, n=3	Ns.	MW P=0.82
		Proband 1	rHSP47+ vs. rHSP47-	213.05(69.75) vs. 72.31(10.98)	n=3, n=3	*	MW P=0.046
		Proband 2	rHSP47+ vs. rHSP47-	146.75(19.41) vs. 37.81(21.27)	n=3, n=3	*	MW P=0.049
		Proband 3	rHSP47+ vs. rHSP47-	51.70(14.33) vs. 14.71(9.73)	n=3, n=3	*	MW P=0.049
		Control vs. Proband 1	rHSP47-	123.65(22.95) vs. 72.31(10.98)	n=3, n=3	**	P=0.017
		Control vs. Proband 2	rHSP47-	123.65(22.95) vs. 37.81(21.27)	n=3, n=3	***	P<0.001
		Control vs. Proband 3	rHSP47-	123.65(22.95) vs. 14.71(9.73)	n=3, n=3	***	P<0.001
		KW= 11.207, p=0.0107					
Figure 5G	Matrix collagen	Control	rHSP47+ vs. rHSP47-	112.02 (3.79) Vs 100.26 (1.58)	n=3, n=3	Ns.	MW P=0.1
		α1(I)G478S	rHSP47+ vs. rHSP47-	94.15 (3.60) Vs 72.84 (1.75)	n=3, n=3	*	MW P=0.04
		α1(I)G667R	rHSP47+ vs. rHSP47-	71.96 (6.21) Vs 63.83 (0.78)	n=3, n=3	Ns.	MW P=0.1
		α1(I)G994D	rHSP47+ vs. rHSP47-	51.68 (3.35) Vs 59.14 (1.02)	n=3, n=3	Ns.	MW P=0.1
		Control vs. α1(I)G478S	rHSP47-	100.26 (1.58) Vs 72.84 (1.75)	n=3, n=3	**	MW P=0.01
		Control vs. α1(I)G667R	rHSP47-	100.26 (1.58) Vs 63.82 (0.78)	n=3, n=3	**	MW P<0.001
		Control vs. α1(I)G994D	rHSP47-	100.26 (1.58) Vs.	n=3, n=3	**	MW P=0.01

59.14 (1.02)						
Figure 6B	Snout-operculum length	rHSP47+ vs. rHSP47-	2.52 (0.40) Vs. 2.45 (0.42)	n=40, n=48	Ns.	MW P=0.4
	Height at eye	rHSP47+ vs. rHSP47-	1.87 (0.38) Vs. 1.86 (0.34)	n=40, n=40	Ns.	MW P=0.8
	Snout-operculum length/ height at eye	rHSP47+ vs. rHSP47-	1.39 (0.16) Vs. 1.35 (0.17)	n=40, n=40	Ns.	MW P=0.4
Figure 6D	Notochord	rHSP47+ vs. rHSP47-	2.17(0.70) vs. 1.85(0.67)	n=41, n=41	*	MW P=0.031
Figure 6E	Ceratohyal bone	rHSP47+ vs. rHSP47-	1.90(0.92) vs. 1.51(0.79)	n=41, n=41	*	MW P=0.039
Figure 6F	Ceratobranchial	rHSP47+ vs. rHSP47-	2.93(0.26) vs. 2.79(0.42)	n=41, n=41	*	MW P=0.049
Figure 6G	Mineral content	rHSP47+ vs. rHSP47-	0.03(0.03) Vs. 0.012(0.02)	n=32, n=32	*	MW P=0.0117