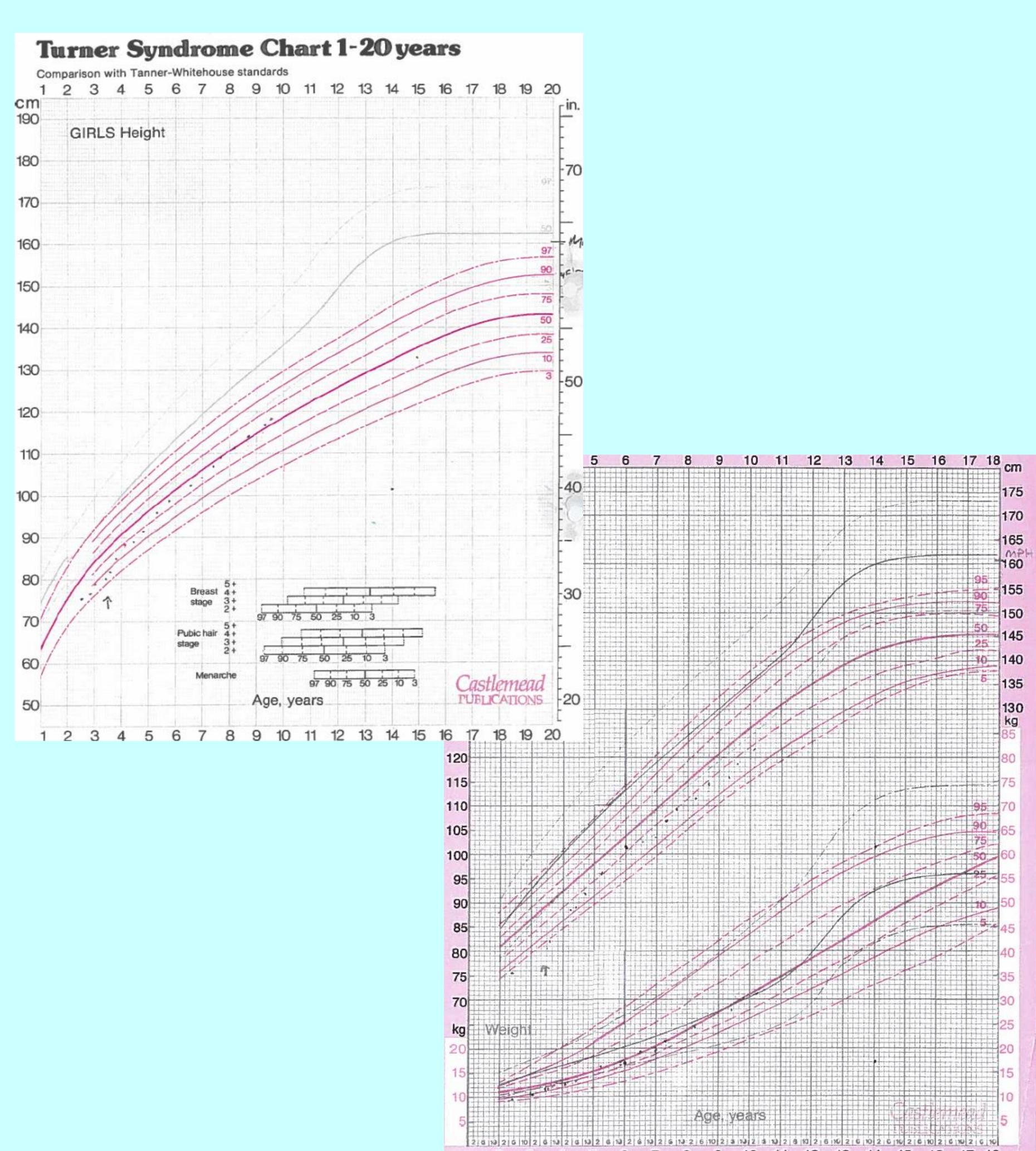
Low dose Growth Hormone using IGF1 dose titration is associated with sustained optimal growth in a child with both Turner and Down Syndrome

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Background: Short stature occurs in both Trisomy 21 and Turner syndrome. This unusual case has a de novo mutation of 47,X,del(X)(p22.3),+21 with clinical features of both syndromes. Growth assessment data and investigations was previously discussed in ESPE2016.

Case: Following growth assessment due to parental concern that her short stature was too short for either syndrome and a falling height velocity of 3.5 cm/year at 2.8 years, the decision was made to start growth hormone (GH) treatment. Low dose GH was started with the aim of reaching Turner syndrome (TS) dose(9.8mg/m²/week). IGF1 levels were closely monitored. Excellent growth response is seen with TS HtSDS -2.08, <5th centile Down's chart improving to TS HtSDS -0.09, just beneath 25th centile Down's chart despite a reduction in GH dose. IGF1 levels pre treatment were good at 171 ng/ml (51-303). Higher IGF1 levels during treatment lead to dose reduction of GH. Unfortunately not all monitoring blood tests were successful and not all blood samples reached the GH reference laboratory. Results and auxology have been tabulated.



						2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18			
	3.12.10	31.10.11	12.11.12	11.11.13	13.8.14	15.6.15	6.5.16	3.11.17	4.5.18
TS HtSDS	-2.08	-1.29	-1.0	-0.57	-0.54	-0.44	-0.32	-0.09	+0.05
Non TS HtSDS	-3.95	-2.2	-2.2	-2.59	-2.6	-2.59	-2.57	-2.59	-2.55
BMI SDS	+0.31	+1.0	+1.07	+0.95	+1.32	+1.41	+1.44	+1.75	+1.46
IGF1	171 ng/ml (51-303)	327 ng/ml (49-289)		49.7 nmol/L (3.3-31.7)	57.9 nmol/L (5.1- 48.2)	48.2 nmol/L (5.1- 48.2)	50.2 nmol/L (5.1- 48.2)	60.9 nmol/L (6.4- 71.9)	
GH (mg/m ² /wk)		8.4	9.2	9.5	5.8	4.4	4.8	6.3	6.4

Conclusion: Dose titration of GH with IGF1 monitoring is necessary in this case to use a lower dose of GH for optimal growth. Parents were pleased with the other non linear growth-related benefits of treatment like improved appetite and weight gain. You can see with increased BMI and age and possible physiological increased GH resistance that GH dosage could be increased again with IGF1 levels in range.







