An Irish regional Study of Paediatric Growth Hormone deficiency (CO-GHD): Classification of Causes and Factors Associated with Persistent GHD .

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BACKGROUND

- The diagnosis of Childhood onset GHD can be challenging.
- Classification of patients according to the underlying cause assist with the prognostic expectations and construction of the transition program when final height is reached.
- The best management approach of adolescents with CO-GHD at the end of growth remains controversial.

OBJECTIVES

- The study aimed to describe all children with CO-GHD in a large regional cohort with clear aetiological classification of children within this cohort.
- To determine the clinical, radiological and growth trajectories including factors affecting final height in each category and predictors of persistent GHD in adulthood.

Methods

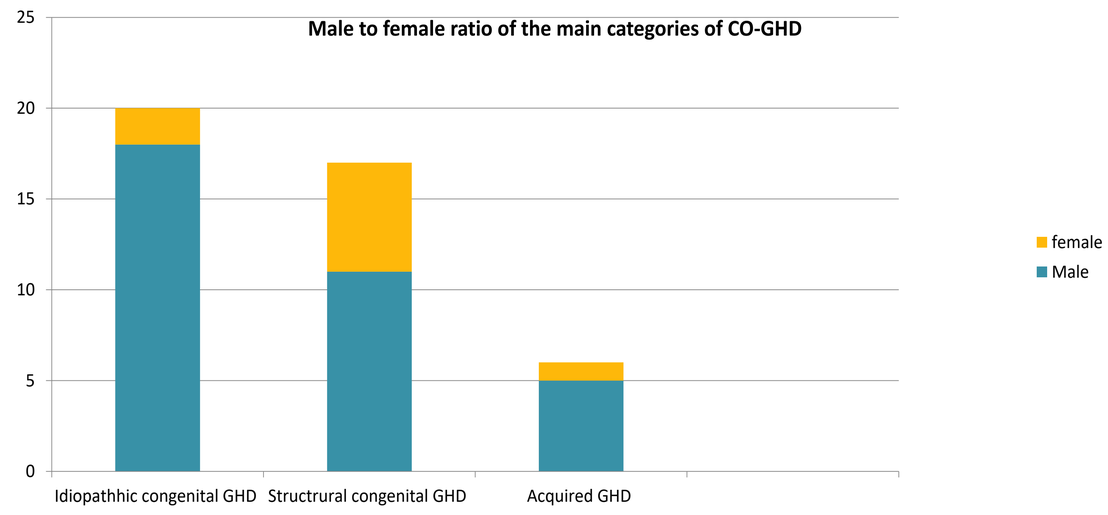
Retrospective cohort study over 2 year (2013- 2015), including all children with CO-GHD and who received recombinant growth hormone treatment rGH treatment – Including children with acquired GHD.

Results

- A total of 43 children fulfilled inclusion criteria of which congenital GHD was identified in 37.
- In congenital GHD 46 % had structural pituitary abnormalities.
- Six children had acquired GHD due to pituitary tumours.
- High male predominance was noted in the 3 main categories, significantly pronounced in in the idiopathic GHD group (9:1)

Diagnosis	N (% of total)
Idiopathic GHD	20 (18.3%)
Congenital GHD due to pituitary Structural defect	15 (16%)
Congenital GHD due to known genetic defect – POU1F1 mutation	2 (2.4%)
Craniopharyngioma	5 (4.9%)
Pituitary macroadenoma	1 (1.2%)

Categories of the cohort of children with CO-GHD-



Clinical features	Idiopathic GHD	Structural Congenital GHD	Acquired GHD	P value
Age at diagnosis	7.6 ± 2.5	4.4 ± 3.8	11± 3.7 SDS	P 0.001
Ht SDS at diagnosis	-2.7 ± 0.9	-2.8 ± 1.07	-1.5± 1.3	p 0.034
HVSDS* at diagnosis	-1.4 ± 1.3	-1.5 ±2.7	-5.1 ± 1.6 SDS	P 0.001
Ht gain SDS-	1.2 ± 0.76	2.3 ±1.6 SD	n/a due to small no	
Ht SDS at transition	-1.2 ± 0.6 SD	-0.86 ± 0.7		

Comparison between at the clinical features at diagnosis and at transition between the 3 main categories

- At final height, 4 of 7 adolescents retested for GHD (57%) exhibited persistent GHD.
- IGF-1 SDS after interruption of treatment < -2 SD correlated with GH status at transition (p=0.04).
- The underlying aetiology was a factor in prediction of GH status at final height, with complex pituitary defects more likely to be associated with persistent GHD (p=0.02).

CONCLUSIONS

- This Irish study revealed novel characteristics such as higher male predominance in congenital idiopathic and acquired GHD due to pituitary tumours.
- A higher percentage of pathological congenital GHD was noted in this cohort compared to the literature.
- New insights on pituitary genetic mutations have emerged during the study, with future implications on the management of GHD at childhood and at the transition to adult care in the affected patients.

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