I MARTEDì DELL'ORDINE

7 GIUGNO 2016

IL BAMBINO CON MALATTIA CRONICA NELLA FASE DI TRANSIZIONE TRA PLS E MMG

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TRANSIZIONE IN ENDOCRINOLOGIA

Transition of adolescents and young adults with endocrine diseases to adult health care

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Wieland Kiess, Prof, Head of the Pediatric Department

Best Practice & Research Clinical Endocrinology & Metabolism 29 (2015) 505–513

Research agenda

- Trials are needed to evaluate the success of different transition modes and patient acceptance.
- Guidelines for the transitional period for different endocrine disorders are lacking.
- Drop out of continuous care at early adult life should be avoided by reassuring that patients have been well received in adults service.
- The way of securing transition success remains unclear

TRANSIZIONE IN ENDOCRINOLOGIA

- **1) Sindrome di Turner**

Sindrome di Turner

Treatment of Turner's syndrome during transition

Aneta Gawlik and **Ewa Malecka-Tendera**

European Journal of Endocrinology
(2014) **170**, R57–R74

Table 1 Turner's syndrome – natural history of associated medical problems.

	Birth/infancy	Preschool	School	Adolescence	Young adulthood
Short stature		→	→	Consequences of GH therapy	Consequences of GH therapy
Cardiovascular disorders	→	→	→	→ Hypertension	→ Hypertension
Otolaryngology problems		→ Otitis media/CHL	→ Otitis media/CHL		→ SNHL
Delayed puberty/ovarian failure				→ HRT	→ HRT/infertility
Renal anomalies	→	→	→	→	→
Obesity/metabolic disorders			→	→	→
Autoimmune diseases				→	→
Behavioural difficulties		→	→ +Specific learning difficulties	→ +Specific learning difficulties	→
Visuo-spatial difficulties					→
Osteoporosis					→
Other features of TS	→	→	→	→	→

→, present; CHL, conductive hearing loss; SNHL, sensorineural hearing loss; HRT, hormonal replacement therapy.

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Table 4 Hormone replacement therapy in Turner's syndrome: suggestions (2).

Age	Suggestions	Route	Dose
10–11	Puberty assessment/FSH/AMH	t/d	Decision about low-dose E_2 (?)
12–13	No puberty/elevated FSH/check AMH	t/d	6.25 μg/day
		p.o.	0.25 mg/day micronised E_2
		i.m.	0.2–0.4 mg/month depo E_2
12.5–15	Gradual increase in E_2 dose over 2 years to adult dose	t/d	Adult: 100–200 μg
		p.o.	Adult: 2–4 mg micronised E_2
			Adult: 20 μg EE_2
			Adult: 1.25–2.5 mg CEE
14–16	After 2 years or when breakthrough bleeding occurs – add cyclic progesterone	p.o.	Adult: 200 mg/day (cyclic)
14–30	Full dose of oestrogen	t/d	Adult: 100–200 μg
		p.o.	Adult: 2–4 mg micronised E_2
			Adult: 20 μg EE_2
			Adult: 1.25–2.5 mg CEE
30–50	Lower dose of oestrogen but providing full protection against osteroporosis		e.g. 0.625 CEE
>50	As in other postmenopausal women		

AMH, anti-Müllerian hormone; E_2, oestradiol; EE_2, ethinyl oestradiol; CEE, conjugated equine oestrogens; t/d, transdermal.

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Sindrome di Turner

Standardized Multidisciplinary Evaluation Yields Significant Previously Undiagnosed Morbidity in Adult Women with Turner Syndrome

J Clin Endocrinol Metab, September 2011,

Kim Freriks, Janneke Timmermans, Catharina C. M. Beerendonk, Chris M. Verhaak, Romana T. Netea-Maier, Barto J. Otten, Didi D. M. Braat, Dominique F. C. M. Smeets, Dirk H. P. M. Kunst, Ad R. M. M. Hermus, and Henri J. L. M. Timmers

TABLE 2. Screening protocol for adults with TS [adapted from Bondy (5), Saenger *et al.* (6), Conway (7)]

Investigation	Once	Every 1–2 yr[a]	Every 3–5 yr[a]
Height, weight, blood pressure, auscultation of the heart		X	
Creatinine, blood urea nitrogen, ASAT, γ-GT, TSH, fT_4, total cholesterol, low-density cholesterol, high-density cholesterol, triglycerides, glucose, HbA1c, urine dipstick analysis		X	
Celiac serology[b]			X
Karyotype	X		
Renal ultrasound	X		
Pelvic ultrasound	X		
Audiogram[c]	X		
Cardiac ultrasound, including electrocardiogram			X
MRI aorta (thoracic and abdominal)			X
Bone mineral density measurement (DEXA)			X

ASAT, Aspartate-aminotransferase; fT_4, free T_4; γ-GT, γ-glutamyl transpeptidase; HbA1c, glycated hemoglobin.

[a] Frequencies are increased when abnormalities are found.

[b] IgA antibodies to tissue transglutaminase and endomysium; in case of IgA deficiency, IgG antibodies were measured.

[c] Repeat at age of 40.

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TABLE 3. Case finding definitions

	Case finding definition
Congenital anomalies of the heart/aorta	Bicuspid aortic valve, coarctation of the aorta, elongation of the transverse aortic arch
Dilation of the aorta	Dilation of the ascending aorta based on BSA corrected values (17, 40) and/or local or fusiform dilation of the descending aorta
Diabetes	Diabetes: random glucose >11.1 mmol/liter or Hba1c ≥6.5 mmol/liter Impaired glucose tolerance: random glucose between 7.8 and 11.1 mmol/liter or HbA1c between 5.7 and 6.4% (41)

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Hypertension	Use of antihypertensive medication or systolic blood pressure >140 mm Hg and/or a diastolic blood pressure >90 mm Hg
Dyslipidemia	TC ≥5.50 mmol/liter and/or HDL-C ≤1.10 mmol/liter and/or LDL-C ≥3.50 mmol/liter and/or TG ≥2.20 mmol/liter (random lipid profiles)
Diminished bone mineral density	Osteoporosis: T-score ≤−2.5 SD; osteopenia: T-score between the −1.0 and −2.5 SD

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Hypothyroidism	Hypothyroidism: TSH above upper reference limit of 4.0 mEq/liter and fT_4 below lower reference limit of 8.0 pmol/liter Subclinical hypothyroidism: TSH >4.0 mEq/liter and fT_4 within normal range (8.0–22.0 pmol/liter)
Structural anomalies of the kidney	Horseshoe kidney, duplication of the collecting system, agenesis, and rotation as TS-specific renal malformations
Liver enzyme elevation	g-GT >35 U/liter and/or ASAT >40 U/liter

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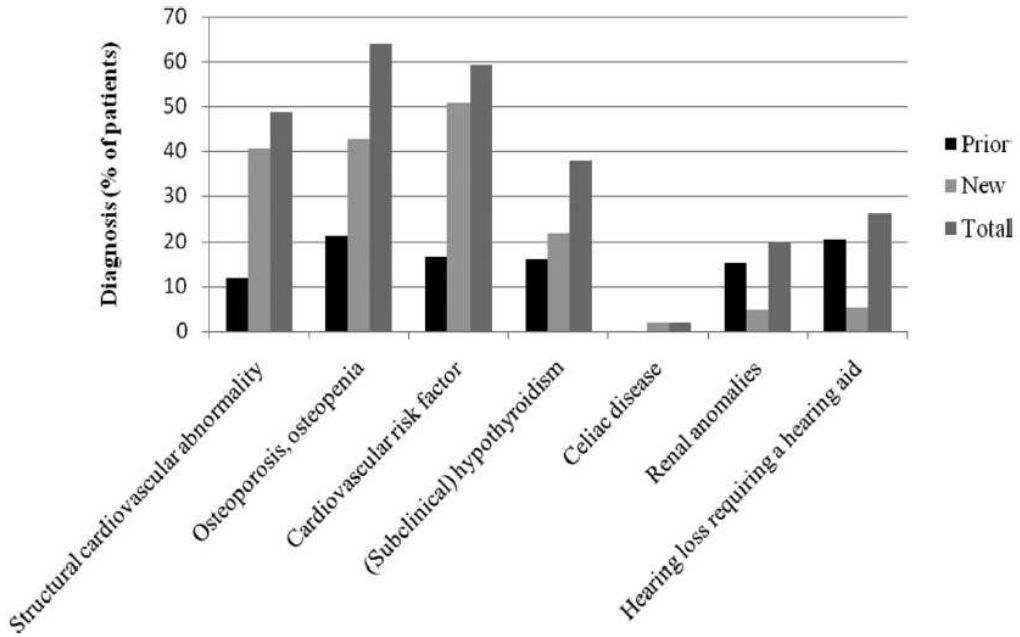

FIG. 1. Frequencies of TS-associated morbidities in 150 TS patients.

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In conclusion, standardized multidisciplinary evaluation of adult women with TS yields significant previously undiagnosed morbidity. TS patients are therefore likely to benefit from a careful transition from pediatric into adult medical care, consisting of a multidisciplinary service with standardized screening for TS-associated morbidity. To what level our approach improves long-term morbidity, mortality, and health-related quality of life remains to be investigated. The actual benefit should also be weighed against the costs of screening and the risk of medicalization.

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Sindrome di Turner

Fertility Preservation in Women with Turner Syndrome: A Comprehensive Review and Practical Guidelines

J Pediatr Adolesc Gynecol xxx (2015) 1–8

Kutluk Oktay MD, PhD [1,2,*], Giuliano Bedoschi MD [1,2], Karen Berkowitz MD [3], Richard Bronson MD [4], Banafsheh Kashani MD [5], Peter McGovern MD [5], Lubna Pal MD [6], Gwendolyn Quinn PhD [7,8], Karen Rubin MD [9]

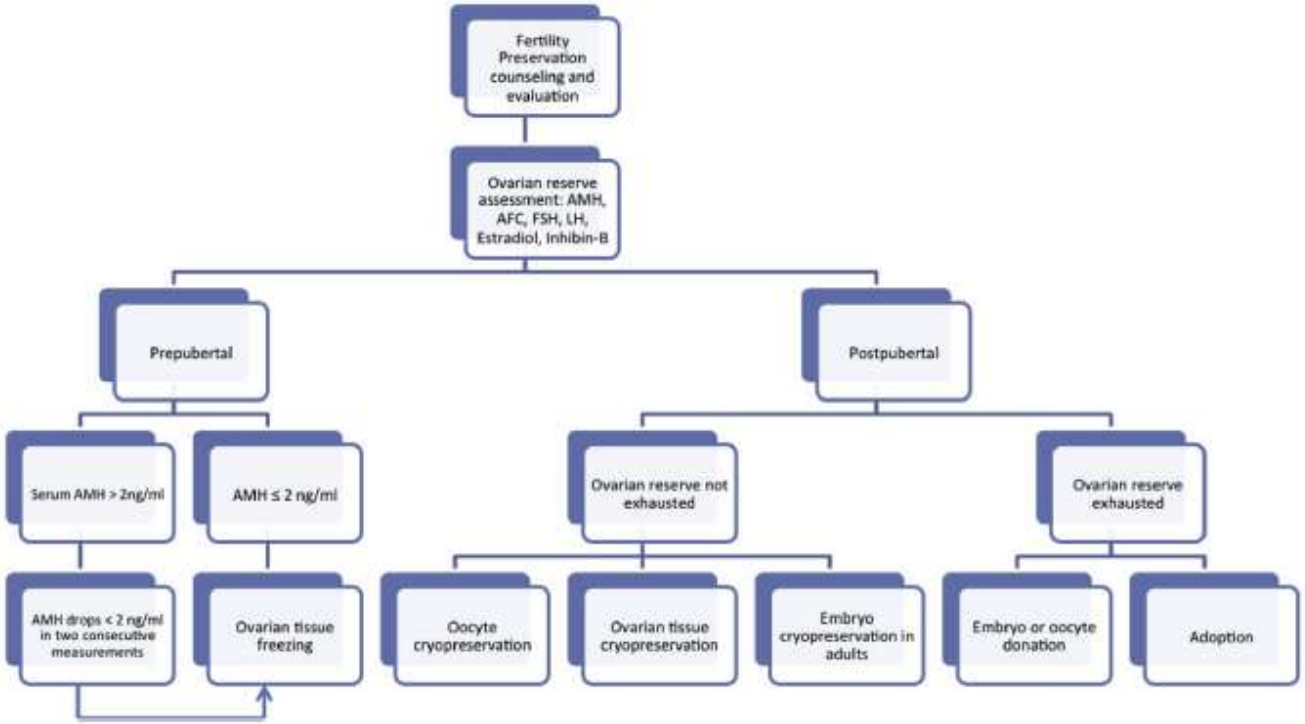

Fig. 1. A proposed algorhythmic approach to decision-making for fertility preservation in females diagnosed with Turner syndrome. For prepubertal girls with sufficient ovarian reserve, expert experience dictates utility of serial serum anti-Müllerian hormone (AMH) assessments to delay intervention to a postpubertal age so that oocyte cryopreservation can be considered. Serum AMH level of less than 2 ng/mL corresponds to levels in the lower quartile for girls aged 5-13 years of age.[23] In postpubertal girls, because the risk of follicle loss is extremely high and can proceed at a fast pace, we recommend fertility preservation regardless of the initial AMH.

Sindrome di Turner

American Journal of Medical Genetics 135A:150–154 (2005)

Gonadoblastoma in Turner Syndrome and Y-Chromosome-Derived Material

Laura Mazzanti,[1]* Alessandro Cicognani,[1] Lilia Baldazzi,[2] Rosalba Bergamaschi,[1] Emanuela Scarano,[1] Simona Strocchi,[1] Annalisa Nicoletti,[2] Francesca Mencarelli,[1] Mariacarla Pittalis,[3] Antonino Forabosco,[4] and Emanuele Cacciari[1]

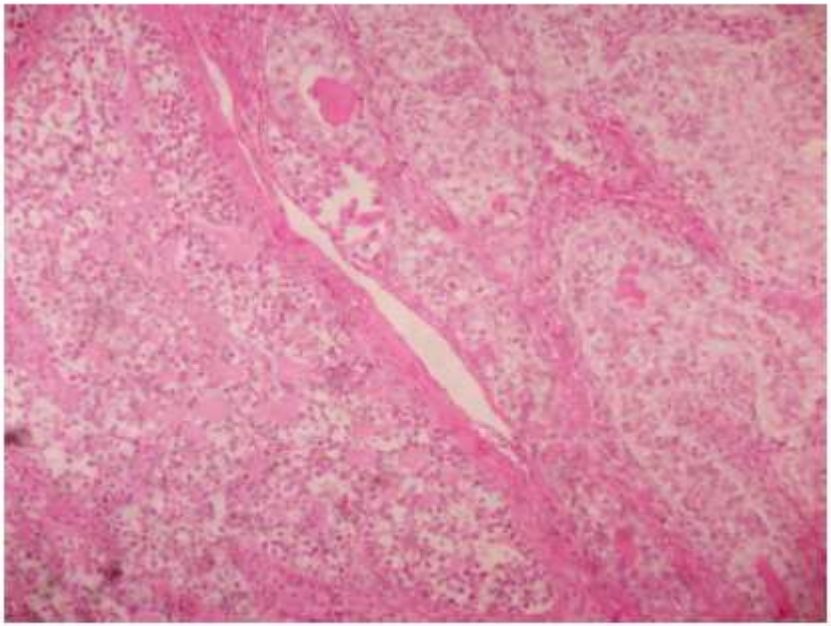

Fig. 1. Microscopic view of the gonadoblastoma (**left**), the immature teratoma and the endodermal sinus carcinoma (**right**) in the left gonad of a patients with UTS (16 years) with Y-mosaicism.

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TABLE II. Three Unselected Series of Patients With Turner's Syndrome Evaluated for Y-Chromosome Material and Gonadoblastoma

	Patients no.	Patients with Y-chromosome material (%)	Gonadectomized patients	Patients with gonadoblastoma (%)	Patients with malignant degeneration (%)
Gravholt et al. [2000]	114	14 (12.2)	10	1/10 (10)	0/1
Alvarez-Nava et al. [2003]	52	4 (8)	4	2/4 (50)	0/2
Our patients	171	14 (8)	12	4/12 (33)	1/4 (25)
Total	337	32 (9.5)	26	7/26 (26.9)	1/7 (14.3)

Sindrome di Turner

Health-care problems of Turner syndrome in the adult woman: a cross sectional study of a Victorian cohort and a case for transition

C. C. Pedreira,[1] R. Hameed,[1] S. Kanumakala[2] and M. Zacharin[1]

Internal Medicine Journal **36** (2006) 54–57

The principal findings of this cross sectional study were of sporadic surveillance and follow up of a group of women at significant risk for ongoing and evolving medical issues, with less than half the cohort being regularly seen for specialist review. Eighty seven percent of the whole cohort had two or more disorders associated with their underlying condition, several members having either been undiagnosed or not treated at the time of this study.

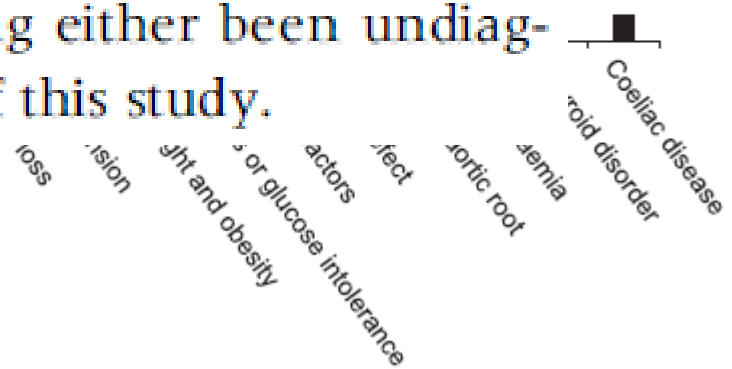


Figure 1 Comorbidities in a cohort of adults with Turner's syndrome.

Sindrome di Turner

Transition in Turner's syndrome

Paul Saenger *

Growth Hormone & IGF Research 14 (2004) S72–S76

To provide a continuum of patient care, medical centers should develop teams with the appropriate expertise in the treatment of Turner's syndrome. These teams should include specialists in the following areas:

- Endocrinologists
- Cardiologists
- Fertility specialists
- Nephrologists
- Audiologists
- Plastic surgeons
- Dentists
- Psychologists

TRANSIZIONE IN ENDOCRINOLOGIA

- 1) Sindrome di Turner
- 2) GHD

Endocrinologi Pediatri

Endocrinologi adulto

GHD

The challenge of growth hormone deficiency diagnosis and treatment during the transition from puberty into adulthood

Elena Inzaghi[1] *and Stefano Cianfarani*[1,2]*

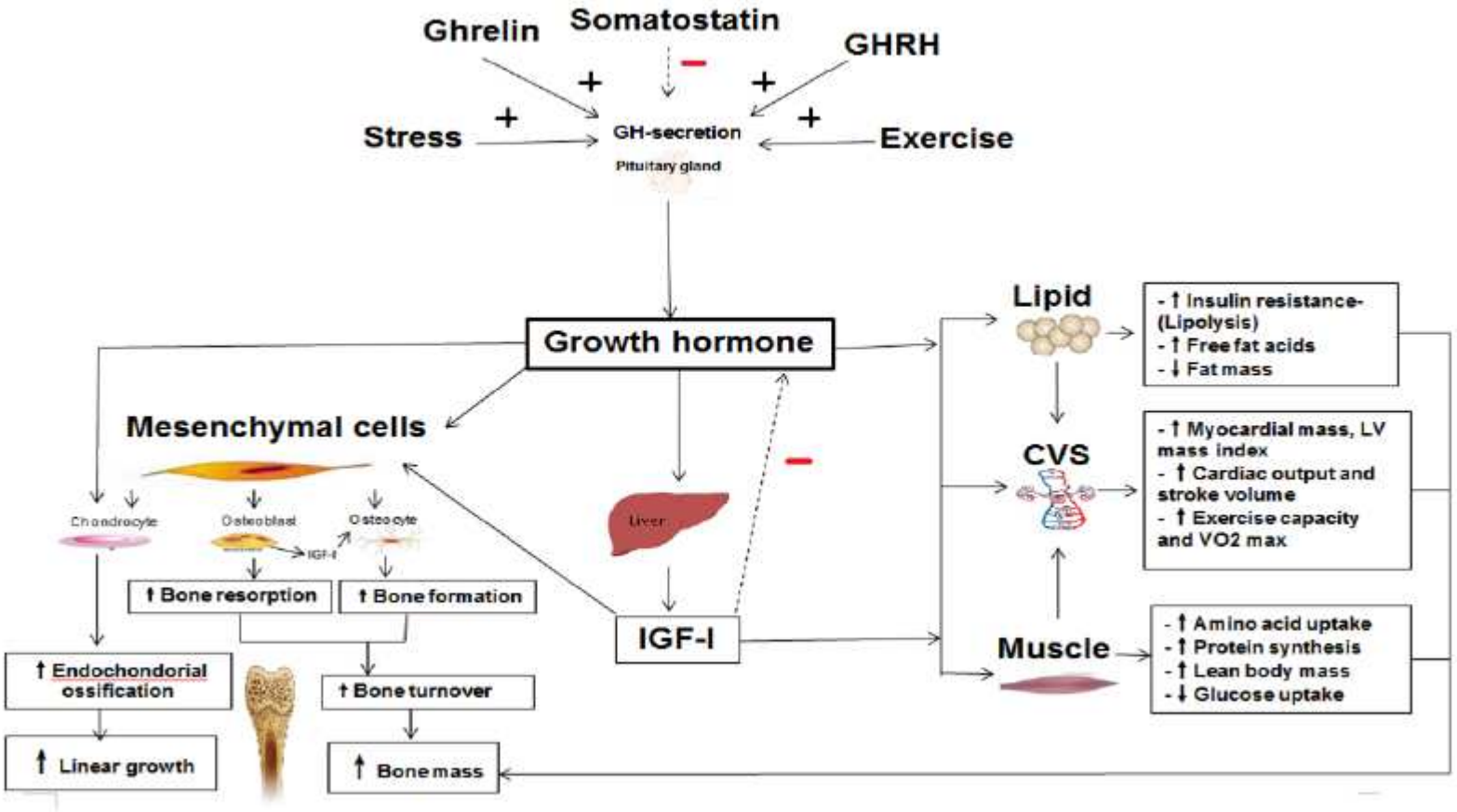
frontiers in ENDOCRINOLOGY

March 2013 | Volume 4 | Article 34 | 1

Table 1 | Features of growth hormone deficiency (GHD) in adult patients.

Features of GHD adult patients
Reduced lean body mass and increased visceral adiposity
Reduced bone mineral density with increased risk of fractures
Reduced IGF-I levels
Decreased muscle strength and exercise capacity
Diminished quality of life (less cognitive function, decreased of well-being)
Abnormal serum lipid profile (increased total cholesterol, LDL cholesterol, triglycerides, lipoprotein A, apolipoprotein B; decreased HDL cholesterol)
Lower cardiac function, impaired left ventricular performance, and increased prevalence of cardiovascular disease

GHD



IL BAMBINO C
CRONICA NELLA FASE DI
TRANSIZIONE TRA PLS E MMG

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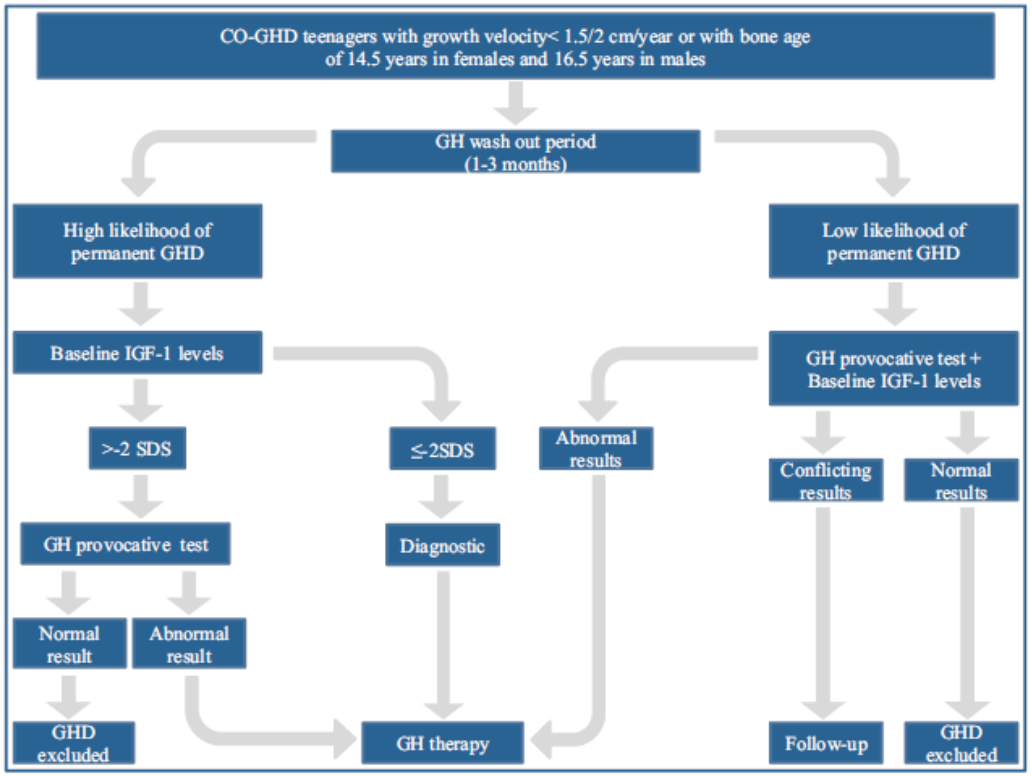

FIGURE 1 | The proposed workup for assessing transition patients with growth hormone deficiency. From a consensus statement issued by the European Society for Pediatric Endocrinology (Clayton et al., 2005).

Clinical Endocrinology (2016) **84**, 708–714

Evaluation of growth hormone stimulation testing in child

Alexander D. Chesover* and Mehul T. Dattani†

All departments stopped GH at least 1 month before retesting, except 3 (6%) departments choosing not to. Specifically, GH was stopped for 1–3 months by 18 departments (38%) and 5 (10%) stated alternatives (0·5, 1·5 and 3 months).

Of those that undertake retesting in the transition period, 19 (56%) use the glucagon test, 16 (47%) insulin, 5 (15%) arginine and 3 (9%) the GHRH–arginine test (missing data, $n = 4$).

Table 3. During the transition period, the number of departments that: Scenario 1: retest certain patient groups. Scenario 2: routinely check IGF-1 in certain patient groups prior to retesting. Scenario 3: still retest certain patient groups after a low IGF-1

		ments	
		rio 2†	Scenario 3‡
		%)	1 (2%)
		3%)	11 (23%)
		5%)	21 (44%)
		0%)	4 (8%)
		%)	4 (8%)
or pituitary disease			
Tumour/surgery/irradiation to hypothalamus or pituitary	8 (17%)	3 (6%)	5 (10%)
All patients	5 (10%)	19 (40%)	6 (13%)
No patients retested	2 (4%)	N/A	N/A
IGF-1 not routinely checked	N/A	9 (19%)	N/A
IGF-1 not routinely checked before retesting	N/A	N/A	4 (8%)
Other	11 (23%)	8 (17%)	12 (25%)

IL BAMBINO CON MALATTIA CRONICA NELLA FASE DI TRANSIZIONE TRA PLS E MMG

Evaluation of growth hormone stimulation testing in children

Alexander D. Chesover* and Mehul T. Dattani†

Clinical Endocrinology (2016) **84**, 708–714

GH 'cut-off' (μg/l)	Number of departments
A	
4	0
6	7 (15%)
6·7	16 (33%)
7	17 (35%)
8	1 (2%)
10	1 (2%)
Other	6 (13%)

GH assay	On-site assay	Referral centre assay
B		
Elecsys Human Growth Hormone Assay (Roche)	4	2
IMMULITE 1000 (Siemens)	2	1
IMMULITE 2000, XPi (Siemens)	13	11
IMMULITE 2500 (Siemens)	2	2
iSYS (IDS)	7	3
Other/Do not Know	2	4

GHD

Documento de consenso del área de conocimiento de Neuroendocrinología de la Sociedad Española de Endocrinología y Nutrición para el abordaje del hipopituitarismo durante la transición

Cristina Álvarez-Escolá[a,*], Eva Fernández-Rodríguez[b], José María Recio-Córdova[c], Ignacio Bernabéu-Morón[b] y Carmen Fajardo-Montañana[d], en representación del Área de Conocimiento de Neuroendocrinología de la Sociedad Española de Endocrinología y Nutrición

Endocrinol Nutr. 2013

1. Recomendamos el tratamiento con GH durante el período de transición, ya que para un déficit similar de GH se han observado diferencias importantes entre CO y AO que pueden deberse a la existencia de períodos sin tratamiento en un momento crucial para la adquisición de una adecuada composición corporal.
2. Recomendamos una colaboración estrecha entre los servicios de endocrinología pediátrica y de adultos con objeto de evaluar de forma conjunta a estos pacientes.
3. Se deben reevaluar todos los casos de déficit de GH, a excepción de las indicaciones pediátricas de tratamiento con GH en niños no deficitarios de GH.
4. La reevaluación debiera realizarse al final del periodo de crecimiento longitudinal.
5. Para la reevaluación, el intervalo sin tratamiento no debiera ser inferior a un mes y el resto de los déficits hormonales deben estar corregidos.
6. Los pacientes con patología orgánica o que presentan alteraciones en la línea media con MPHD o los que presentan IGHD con mutaciones en genes que influyen en el desarrollo de la hipófisis o en la expresión del gen de la GH no precisan reevaluación y el tratamiento podría mantenerse ajustando dosis.

GHD

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Endocrinol Nutr. 2013

7. Los pacientes con MPHD idiopático o adquirido e IGHD adquirido de etiología desconocida o bien con antecedentes de tumor, cirugía o irradiación hipofisaria, deben reevaluarse tras un mes sin tratamiento, y si la IGF-1 fuera normal, realizar un test de estímulo.
8. Los casos con IGHD con hipófisis normal y sin antecedentes de interés deben reevaluarse tras al menos un mes sin tratamiento y realizar en todos los casos test de estímulo.
9. Por el momento recomendamos la realización del test de hipoglucemia insulínica, ya que sigue considerándose el método de referencia. En los casos en que estuviera contraindicada, podría utilizarse el *test* de glucagón.
10. Los test de GHRH + arginina o GHRH + GHRP6 requieren la integridad del eje hipotálamo-hipofisario. El test de GHRH + arginina tiene la ventaja de su escasa variabilidad individual y que tiene diferente punto de corte según el IMC.
11. Durante la transición debiera considerarse un punto de corte de GH inferior a 5,1 μg/l con insulina, que equivale a un valor menor de 4,15 μg/l con GHRH + arginina.

GHD

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Endocrinol Nutr. 2013

12. La determinación de IGF-1 puede ayudarnos. Hay que tener en cuenta, que tras la suspensión del tratamiento con GH, puede tardar en alcanzar su nivel «basal». El punto de corte de IGF-1 se establece en menos de −2 DE (aproximadamente 100 μg/dl)[2]. Se deben recordar las causas de niveles falsamente bajos.
13. La dosis inicial debiera ser de 0,2 a 0,5 mg/día. Se deben recordar las interferencias con otros tratamientos hormonales y no olvidar que el cambio de dosis de GH puede hacer necesario el ajuste de las dosis de los restantes tratamientos sustitutivos.
14. Debiera determinarse IGF-1 a las 6 semanas del inicio del tratamiento para ajustarlo con objeto de mantener IGF-1 en el percentil 25 a percentil 75 del rango normal. Una vez ajustado, debiera determinarse IGF-1 cada 6 meses. Para evaluar cualquier cambio de dosis debería medirse IGF-1 al menos 6 semanas después. Anualmente se evaluará peso, IMC, cociente cintura/cadera, tensión arterial, pulso y test de calidad de vida. Cada 2 a 5 años se realizará perfil lipídico y densitometría, valorando el T-score en la década de los 20 años, y T y Z score posteriormente.

GRAZIE PER L'ATTENZIONE