

Gonadotropin Levels And Rate Of Testosterone Supplementation In Adolescents With Klinefelter Syndrome

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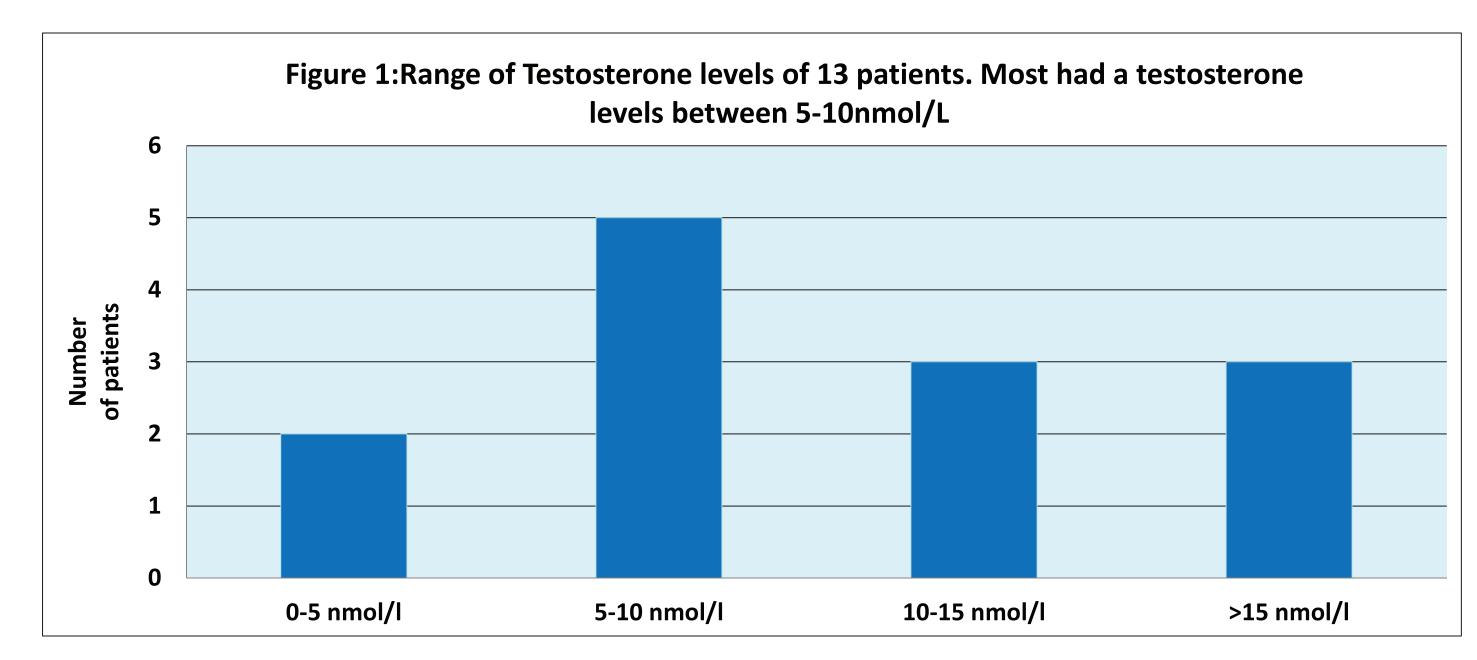
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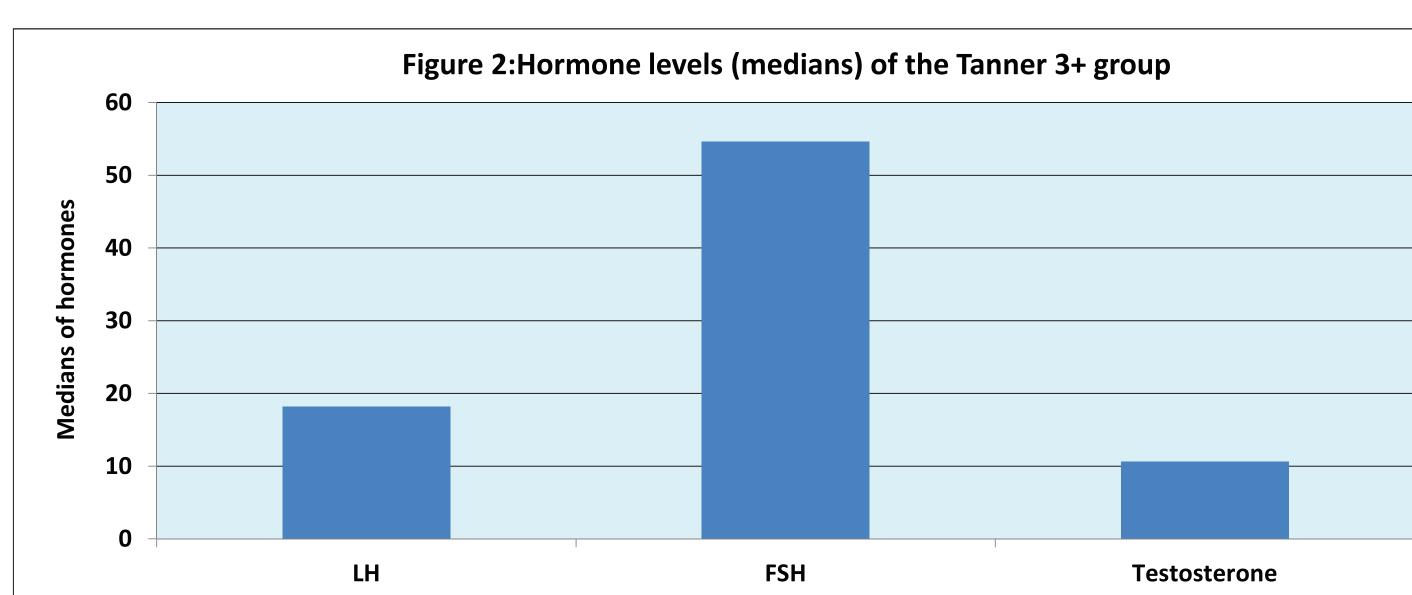
Introduction

Klinefelter syndrome (XXY) is characterised by sex hormone aneuploidy. It is one the most common causes of primary hypogonadism, impaired spermatogenesis and testosterone deficiency. It occurs in up to 1 out of 500 to 700 phenotypic males. Approximately 25% of the individuals are diagnosed in childhood. The hallmark of the condition are small testicular volumes and azoospermia. The testosterone levels are often normal at the expense of elevated gonadotrophins (LH, FSH).

Results

- Seventeen adolescents of pubertal age were identified.
- The age range was 11.7 to 17.1 years at the time of referral
- Four out of seventeen patients presented with gynaecomastia.
- All but one were in puberty with the majority (13/17) being between Tanner stage 3 to 5. Two patients did not have Tanner stage recorded (Table 1).
- The range of testosterone levels are shown in Figure 1.
- Gonadotrophins were not elevated in adolescents who were at Tanner stage 1 or 2.
- In those with Tanner stage 3+, median LH was 18.2 IU/L, FSH 54.6 IU/L and testosterone 10.6 nmol/L. (Figure 2).





• 12 boys with Tanner stage G3+ had testosterone levels done with a range from 4 - 20.1 (median: 10.7 nmol/L). Nine out of twelve had a testosterone level in the lower third of the adult range <15nmol/L.

Aim

- Retrospectively review the investigations and management of puberty of all adolescents with XXY born from 1999 in large tertiary hospital.
- Document the gonadotrophins, testosterone levels and need for additional testosterone
- 3 out of 17 adolescents required testosterone treatment. The median start age of treatment being 15.9 years. All had a testosterone in the lower normal range with a median of 9.8nmol/L, LH of 15.9 and FSH of 68.2 IU/L. (Figure 3)

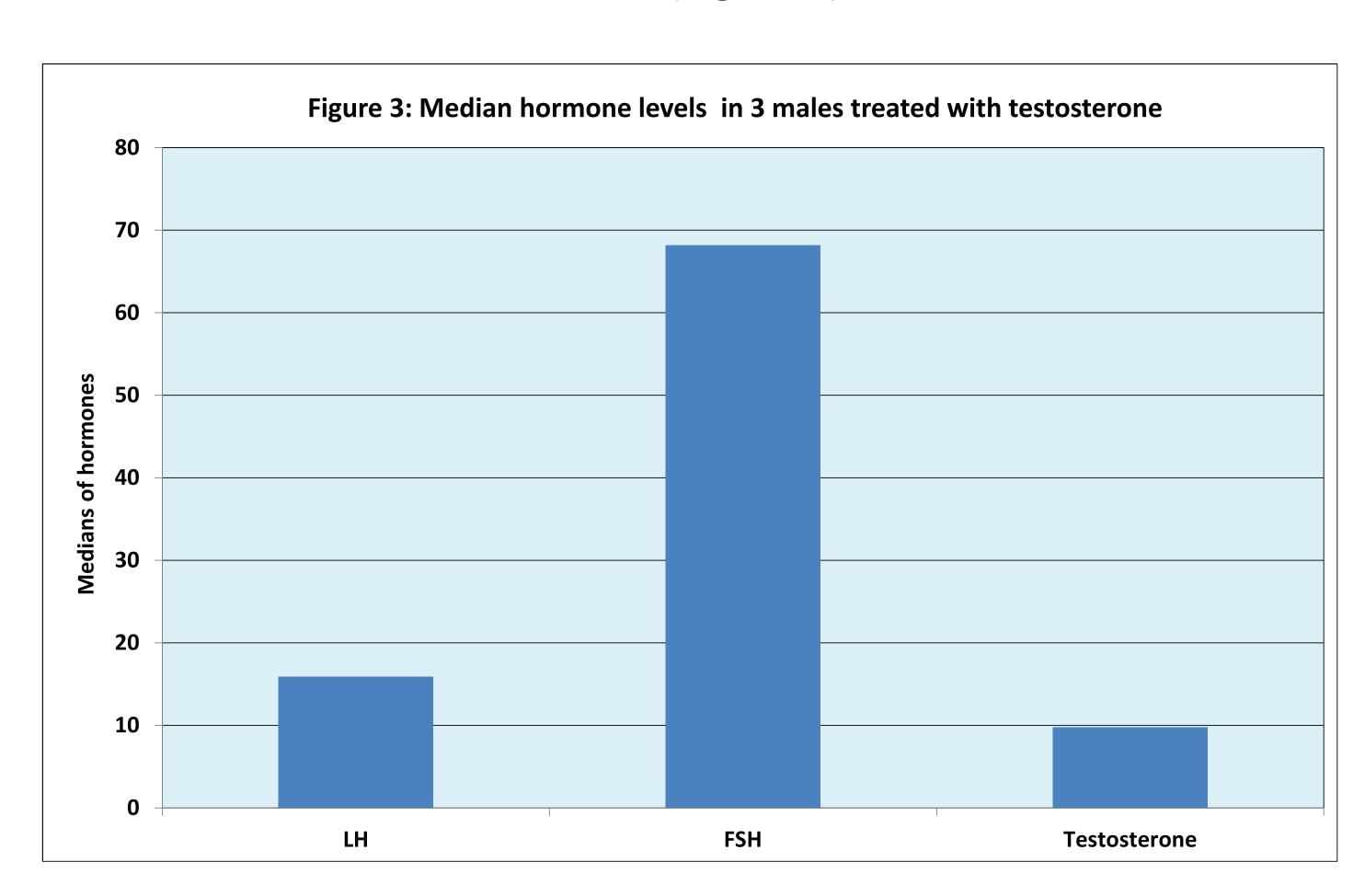


Table 1: Pubertal stage, LH, FSH and testosterone levels in 17 patients

Patient	Decimal age (years)	Tanner stage	LH (IU/L)	FSH (IU/L)	Testo (nmol/L)
1	11.7	G2 P2 A1	0.2	n/a	0.7
2	13.8	G3, P3, A2	17.6	57.4	4
3	16.0	G4 P4	13.8	68.2	6.7
4	15.1	G5 P4 A2	16.4	32.9	7.3
5	16.3	G3, P3, A3	23.6	31.6	8.4
6	17.1	G4 P5 A2	15.9	36.8	9.8
7	15.4	G5 P4	4.6	18.1	9.9
8	14.9	G5 P5 A2	30.9	98.1	11.4
9	15.4	G4 P4 A1	18.8	51.8	11.6
10	13.9	G3 P3 A3	37	80.4	14.6
11	16.8	G4 P4 A3	25	67.5	17.1
12	14.8	G4 P4 A3	10.4	44.6	17.4
13	14.1	G4 P5 A2	26.7	99.2	20.1
14	n/a	G4 P4 A2	n/a	n/a	n/a
15	n/a	n/a	n/a	n/a	n/a
16	n/a	G1P1A1	n/a	n/a	n/a
17	n/a	n/a	n/a	n/a	n/a

Conclusions

Hypogonadism in XXY is common. Three out of thirteen boys who had blood tests done required testosterone treatment. They rarely get testosterone level above the mid-adult range. Teenagers should continue to have surveillance in adult years.

References:

- 1. Groth K. Klinefelter syndrome-A clinical update. J Clin Endocrinol Metab, January 2013, 98(1):20 –30
- 2. Klinefelter syndrome. https://ghr.nlm.nih.gov/condition/klinefelter-syndrome. Published 07/08/2017, Accessed last 13/08/2018

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