

KALLMANN SYNDROME - TREATMENT IN EARLY CHILDHOOD

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Kallmann syndrome (KS) is a neuronal migration disorder characterised by hypogonadotrophic hypogonadism and anosmia or hyposmia. Micropenis and bilateral kryptorchism are the most striking features in early childhood. Intracerebral migration of GnRH neurons is arrested in KS resulting in GnRH deficiency followed by different degrees of luteinizing hormone (LH) and follicle stimulating hormone (FSH) deficiencies resulting in a hypogonadic phenotype.

We report our experience of KS in diagnostics, hormonal treatment and therapeutic results in the first year of life. The literature about KS in early childhood is reviewed. Our cases were referred to us because of extreme hypogonadism and kryptorchism combined with micropenis. Ultrasound of the inguinal region and abdomen was performed. Moreover abdominal MRI was performed. Cranial MRI was used in diagnosing agenesis or hypotrophy of the olfactory sulci. Hormone status included FSH, LH and testosterone. Chromosomal diagnostics always revealed normal 46XY. Treatment consisted of stimulation with HCG and finally substitution of testosterone. Gonadal descensus in the first year of life could be achieved in 50% of the cases. Penis enlargement was satisfactory and growth retardation was compensated.

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