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Case Series - VanWykGrumbachs syndrome in North Karnataka.

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Back-ground: Juvenile hypothyroidism in young females with precocious puberty and multicystic ovaries was first described by Van Wyk and Grumbach in 1960. The exact prevalence of the syndrome is unknown and only sporadic case reports have been reported in the literature since then.

Materials and Methods: The present case series on this rare syndrome is an observatory and descriptive data of three cases of Van WykGrumbach syndrome who were referred to our center for evaluation of precocious puberty.

Results: The mean age of presentation was 9 year 1 month. Two cases presented with menarche as the main complaint, one with thelarche and menarche. On examination all had short stature (height < 3rd centile, weight < 3rd centile), delay of bone age by more than 3 years with high TSH, low T₃ and T₄ at diagnosis. Prepubertal LH with high FSH confirmed the clinical suspicion. USG abdomen showed bilateral multicystic ovaries in two cases and one had only right-sided bulky ovary. One of the patients' MRI showed a Pituitary macroadenoma. MRI wasn't done in the other two cases due to financial constraints.

Conclusions: Precocious puberty, short stature, juvenile primary hypothyroidism and delayed bone age can be taken as a clinical quartet in diagnosing VanWykGrumbach and will always remain a diagnosis of exclusion. Levothyroxine is the treatment of choice and it normalizes all the other hormone levels post replacement along with anatomical changes (shrinking of the ovarian cysts and regression of the pituitary macroadenoma). Regular monitoring with thyroid function tests to maintain euthyroidism is of at most importance to provide a good quality of life.

Key Words – Van WykGrumbach, Precocious Puberty, Primary hypothyroidism

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