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Original Research Paper

Pediatrics

A CASE REPORT OF PARTIAL CONGENITAL HYPOPITUITARISM

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ABSTRACT Where hypopituitarism is common but congenital hypopituitarism is rare. Severe prenatal deficiency of GH as occur in congenital hypopituitarism has little effect on fetal growth. Prenatal and congenital deficiency can reduce the size of male penis especially when gonadotropins are also deficient. Severe GH deficiency in early childhood also result in slower muscular development so that gross motor mile stone such as standing ,walking and jumping may be delayed. Bone age is helpful as it is usually delayed in patient with congenital Growth hormone deficiency.

Incidence: Between 1 in 4000 and 1 in 10000 live births.

KEYWORDS : Congenital Panhypopituitarism, Growth Hormone Deficiency, Development Delay

INTRODUCTION:

Pituitary gland is responsible for the production and secretion of various hormones that play a vital role in the normal growth and development of a child. It consist of anterior and posterior lobe. Anterior lobe produces Growth hormone(GH),Thyroid stimulating hormone(TSH),Luteinizing hormone(LH),Follicle stimulating hormone(FSH),Adrenocorticotropic hormone(ACTH),Prolactin whereas posterior lobe produce Antidiuretic hormone(ADH) and oxytocin. They are in turn regulated by the hypothalamus that either stimulate or inhibit the production of different pituitary hormones.

Hypopituitarism is a condition in which there is underproduction of growth hormone alone or in combination with other pituitary hormones. Multiple pituitary hormone deficiency(MPHD) has 2 types .One is genetic in which sequentially expressed transcription, activation factor direct differentiation & proliferation of pituitary cell type. Mutation in this leads to different MPHD. In Acquired variety any lesion that damage hypothalamus , pituitary stalk or anterior pituitary.

Examples: Craniopharyngioma, CNSgerminoma, Eosinophillic, granuloma, TB, Sarcoidosis, Toxoplasmosis, Meningitis. Growth hormone (GH) promotes linear growth in children by stimulating cartilage growth, particularly at the epiphyseal plate. In addition, GH increases lean body mass and bone mass and reduces fat mass, while increasing plasma and liver lipid content.

CASE REPORT:

A 1 year old male child with weight of 4.9 kg admitted at tertiary care hospital for chief complain of not gaining weight and developmental delay. The Patient has an uneventful birth history with birth weight of 2300 gm. At the time of admission patient was not able to stand with support, with immature pincer grasp , speaks only monosyllable words, smiles at mirror image. On genital examination small phallus, hypo pigmented scrotum was there. X ray wrist was done which shows bone age of 4 months. 2D ECHO and MRI brain were done which was normal. Hormonal evaluation was done , tabulated below. Endocrinologist opinion was taken and they diagnosed it as partial congenital hypopituitarism and advised to start Injection GH 0.5 IU subcutaneous on daily basis. Patient had taken this treatment for 6 months till now. Patient is in regular follow up for growth and development monitoring.

Investigations	Normal value	Before treatment at 1 year of age	After treatment at 1.5 years of age
Human growth hormone	0-3 ng/ml	<0.05	0.12
ACTH	3.6-60 pg/ml	7.03	9.32
Insulin like growth factor-1	55-327 ng/ml	<15	<15
Prolactin	2.1-17.7 ng/ml	<0.32	1.1
Cortisol(random)	3.7-19.4 ug/dl	17.85	3.96
FSH	1.4-18.1mIU/ml	1.25	0.54
LH	1.14-8.75mIU/ml	<0.07	<0.07
T3	0.6-1.81ng/ml	0.68	0.82
T4	4.5-12.6 ug/dl	1.4	1.9
TSH	0.35-5.5 uIU/ml	<0.01	0.40

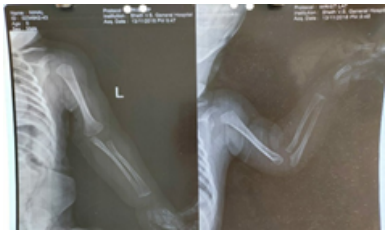
After 6 months of treatment patient is now able to run,feed himself with spillage, speaks 3-4 words with meaning showing improvement over growth and development.

	Before treatment at 1 year of age	After treatment at 1.5 years of age
Height	54cm	58cm
Weight	4.9 kg	9.2kg
Head circumference	41 cm	43cm

- Height velocity is monitored at three- to six-month intervals (algorithm 1); the goal for treatment is to attain the 75th percentile for height velocity for age during catch-up growth. We also monitor IGF-1 levels every 6 to 12 months

CONCLUSION:

Raising awareness about the cause and early initiation of treatment is important because unrecognized pituitary dysfunction significantly affect the physical and psychological well-being. High index of suspicion coupled with timely and appropriate hormonal assay are essential for the management. It needs to be diagnosed and treated early to prevent associated paediatric mortality and morbidity. Supervision, care and regular growth monitoring by paediatrician is crucial along with endocrinologist support.



X ray wrist AP and lateral view suggestive of bone age of 4 months

At 1 year of age **At 1.5 yr of age**