ABSTRAK

Latar belakang

Hipopituitari merupakan kegagalan parsial atau total hormon hipofisis anterior dan atau posterior. Secara global, insidensi hipopituitarisme berkisar 11 hingga 42 kasus baru tiap 1 juta penduduk per tahunnya. Penyebab hipopituitari bisa kongenital atau didapat.[1,2] Manifestasi klinis hipopituitari tergantung pada tingkat dan derajat keparahan defisiensi hormon, durasi penyakit serta onset. Manajemen utama hipopituitarisme pada prinsipnya adalah dengan penggantian hormon target yang kurang. *Childhood Onset Multiple Pituitary Hormone Deficiency (COMPHD)* memiliki dampak pada semua aspek perkembangan somatik, efek patofisiologi dari defisiensi hormon yang spesifik, dan gangguan akan berlangsung seumur hidup, sehingga peran serta semua pihak sangat diperlukan dalam manajemen *COMPHD* secara menyeluruh dan berkelanjutan.[2]

Kasus

Sebuah kasus longitudinal remaja laki-laki berusia 15 tahun telah terdiagnosis *Multiple Pituitary Hormone Deficiency (COMPHD)* sejak usia 8 tahun. Pada pemeriksaan fisik didapatkan berat badan 30 kg, tinggi badan 133,2 cm (HAZ score : -4.91) severely stunted, pemeriksaan status tanner sesuai G2P2A1. Hasil pemeriksaan IQ dalam batas normal, kortisol rendah (< 0,8 µg/dl), IGF-1rendah (<25 ng/ml), FT4 rendah (0,59 ng/dL), TSH meningkat (4,45 µIU/ml), hasil tes stimulasi GH rendah (0,07; 0,1; 0,14; 0,15; 0,13 ng/ml). Hasil pemeriksaan kadar hormon sex steroid pre-pubertal, didapatkan LH, FSH, dan testosteron rendah, menunjukkan adanya defisiensi gonadotropin dan hormon seks steroid. Hasil pemeriksaan USG abdomen dan skrotal, serta MRI kepala dalam batas normal. Hasil bone age Greulich foto manus dextra et sinistra (usia kronologis anak 15 tahun 6 bulan) menunjukkan gambaran sesuai usia 13 tahun 6 bulan berdasar Greulich Radiographic atlas of skeletal development of the hand and wrist. Anak didiagnosis dengan *Multiple Pituitary Hormone Deficiency (COMPHD)*, dan mendapatkan *replacement hormone therapy* (RHT) dengan Growth Hormone, Levothyroxine, dan injeksi testosteron. Dilakukan pemantauan dan evaluasi hormon berkala.

Kesimpulan

Replacement Hormone Therapy (RHT) dengan Growth hormone, levothyroxine, hidrokortison akan memberikan perbaikan pertumbuhan linear pada pasien MPHD. Pemberian injeksi testosteron untuk menginduksi pubertal yang akan memacu *growth spurt* akan lebih efektif jika disertai pemberian *Growth hormone*.

Kata kunci

Hipopituitari, growth hormone, tiroid, kortisol, perawakan pendek, mikropenis, delay pubertas

ABSTRACT

Background

Hypopituitarism is a partial or total failure of anterior and/or posterior pituitary hormones. Globally, the incidence of hypopituitarism ranges from 11 to 42 new cases per 1 million population per year. Hypopituitary causes can be congenital or acquired.[1,2] Clinical manifestations of hypopituitarism depend on the degree and severity of hormone deficiency, duration of disease and onset. The primary management of hypopituitarism is principally replacement of the deficient target hormone. Childhood Onset Multiple
Pituitary Hormone Deficiency (COMPHD) has an impact on all aspects of somatic development, the pathophysiological effects of specific hormone deficiency, and the disorder will last a lifetime, so the participation of all parties is indispensable in the comprehensive and sustainable management of COMPHD.[2]

Case Presentation

A longitudinal case of a 15-year-old boy who had been diagnosed with Multiple Pituitary Hormone Deficiency (COMPHD) since the age of 8 years old. On physical examination, the body weight was 30 kg, the body height was 133.2 cm (HAZ score: -4.91, severely stunted), the Tanner status was G2P2A1. The IQ test results were within normal limits. The hormone examination result were low cortisol (< 0.8 g/dl), low IGF-1 (<25 ng/ml), low FT4 (0.59 ng/dL), increased TSH (4.45 IU/ml), and low GH stimulation test (0.07;
0.1; 0.14; 0.15; 0.13 ng/ml). The pre-pubertal sex steroid hormone levels test revealed low of LH, FSH, and testosterone, indicating a deficiency of gonadotropins and steroid sex hormones. The abdominal and scrotal ultrasound examination and head MRI were within normal limits. The results of Greulich's bone age photo of manus (children's chronological age was 15 years and 6 months old) according to the age of 13 years and 6 months based on Greulich Radiographic atlas of skeletal development of the hand and wrist. The child was diagnosed with Multiple Pituitary Hormone Deficiency (COMPHD), and received replacement hormone therapy (RHT) with Growth Hormone, Levothyroxine, hydrocortisone, and testosterone injections. Periodic monitoring and evaluation of hormones is carried out.

Conclusion

Replacement Hormone Therapy (RHT) with Growth hormone, levothyroxine, hydrocortisone will provide linear growth improvement in MPHD patients. Giving testosterone injections to induce puberty by stimulating growth spurts will be more effective if accompanied by growth hormone administration.

Keywords

Hypopituitarism, growth hormone, thyroid, cortisol, short stature, micropenis,
delayed puberty