Deteksi dini keterlambatan pubertas pasien transfusion dependent thalassemia dengan mri t2* hipofisis: hubungannya dengan profil besi, hormon gonadotropin dan seks steroid = Early detection of delayed puberty in transfusion dependent thalassemia patients with mri t2* hypophysis: correlation with iron profile, gonadotropine and sex steroid hormones

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Abstrak

Latar belakang: Thalassemia adalah kelainan sel darah merah berupa gangguan sintesis hemoglobin (Hb) yang diturunkan secara autosomal resesif. Transfusi PRC pada pasien transfussion-dependent thalassemia (TDT) menyebabkan akumulasi besi pada berbagai organ, salah satunya kelenjar endokrin. Hal tersebut akan menyebabkan hemosiderosis di hipofisis anterior dan keterlambatan pubertas.

Tujuan: Mendeteksi keterlambatan pubertas menggunakan MRI T2* hipofisis di poliklinik Thalassemia Departemen Ilmu Kesehatan Anak Rumah Sakit Cipto Mangunkusumo Jakarta.

Metode: Pemeriksaan MRI dilakukan menggunakan MRI Avanto 1,5 Tesla, sekuen T2 SE. Penghitungan nilai MRI T2* hipofisis menggunakan perangkat lunak CMR Tools TM yang berfungsi menghitung deposit besi pada organ. Dilakukan pemeriksaan kadar feritin serum, FSH, LH testosteron (lelaki), dan estradiol (perempuan) menggunakan electro-chemiluminscence immunoassay (ECLIA). Hubungan antara MRI T2* hipofisis dengan kadar feritin serum, saturasi transferin, FSH, LH, testosteron, dan estradiol dianalisis menggunakan analisis korelasi Pearson.

Hasil: Sebanyak 56 pasien TDT dibagi menjadi 27 subyek usia prapubertas, 14 subyek pubertas normal dan 15 subyek pubertas terlambat. Hasil menunjukkan nilai MRI T2* hipofisis pasien TDT pubertas normal lebih tinggi secara signifikan dibandingkan pubertas terlambat (p=0,000). Terdapat hubungan bermakna antara nilai MRI T2* hipofisis dengan feritin serum (r = -0,502) dan tidak ada hubungan antara MRI T2* hipofisis dengan saturasi transferin, FSH, LH, testosteron, dan estradiol.

Simpulan: Nilai MRI T2* hipofisis TDT pubertas normal lebih tinggi dibandingkan pubertas terlambat.
.....Background: Thalassemia is an autosomal recessive red blood cells disorder characterized by abnormal hemoglobin (Hb) synthesis. PRC transfusion to transfussion-dependent thalassemia (TDT) patients results in iron accumulation in organs, including endocrine system. It further cause hemosiderosis at anterior hypophysis which leads to delayed puberty.

Objective: To detect patients with delayed puberty using MRI T2* in Thalassemia Clinic Department of Pediatrics Cipto Mangunkusumo Hospital Jakarta.

Method: MRI was assesed with MRI Avanto 1,5 Tesla,T2 SE sequence, whilst T2* hypophysis result being

done with CMR Tools TM software, which aims to measure iron deposit. Levels of ferritin serum, FSH, LH, testosterone (male subjects), and estradiol (female subjects) were observed with electro-chemiluminescence immunoassay (ECLIA). Correlation between MRI T2* hypophysis with levels of ferritin serum, transferrin saturation, FSH, LH, testosterone, and estradiol were analyzed with Pearson correlation analysis.

Results: 56 TDT patients were consist of 27 prepuberty subjects, 14 normal puberty subjects, and 15 delayed puberty subjects. Results showed that MRI T2* hypophysis of normal puberty TDT patients was significantly higher compared to delayed puberty patients ($p = 0,000$). There was significant correlation between MRI T2* hypophysis with ferritin serum ($r = -0,502$). Meanwhile, there was no significant correlation between MRI T2* hypophysis with transferrin saturation, FSH, LH, testosterone, and estradiol.

Conclusions: Pituitary MRI T2* of TDT patients higher in normal group than that in delayed puberty group.