

**REPLY**

The above situation described by the author is an interesting and often faced dilemma in pediatric nephrology practice. Since this child has already received 6 weeks of daily steroids and went into remission, the relapse should be technically treated as the first relapse. Most regimens for treatment of initial episode have recommended 4-6 weeks of daily steroids followed by alternate day therapy for another 6 weeks only, as longer durations predispose to more adverse effects [1,2]. We should treat this episode as first relapse and give the child daily prednisolone (2mg/kg/d) till 3 days of remission and then continue on alternate day (1.5 mg/kg/d) of oral prednisolone for another 4 weeks. This means that the child would receive at least another 5-6 weeks of steroids and she had already missed 6 weeks of alternate day steroids during the treatment of initial episode. Even if we consider this episode as continuum of the initial episode the child would still merit 6 weeks of alternate day steroid therapy that she had missed. However since the child

relapsed after gaining remission it should be labeled as a relapse. The definition of relapse as per the guideline is "Urine albumin 3+ or 4+ (or proteinuria >40 mg/m<sup>2</sup>/h) for 3 consecutive early morning specimens, having been in remission previously" [1]. The definition of first relapse or subsequent relapses is not any different. The subsequent treatment of this child would be decided by the disease course on follow-up.

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**REFERENCES**

1. Consensus Statement of Management of Steroid Sensitive Nephrotic Syndrome. Indian Pediatric Nephrology Group, Indian Academy of Pediatrics. Indian Pediatr. 2008;45:203-14.
2. Lombel RM., Gipson DS, Hodson EM. Treatment of steroid-sensitive nephrotic syndrome: new guidelines from KDIGO. *Pediatr Nephrol.* 2013;28:415-26.

## Diabetes Monitoring in Hemoglobinopathies

A-10-year-old boy was recently diagnosed as type I diabetes mellitus. As a part of work up, an HbA1c (glycosylated hemoglobin) was sought but could not be done due to presence of abnormal hemoglobin, later confirmed as HbE trait.

In our experience, we note an increasing number of children with abnormal hemoglobin and diabetes. The Diabetes Control and Complications Trial (DCCT) and the United Kingdom Prospective Diabetes Study (UKPDS) demonstrated conclusively that risks for complications are related directly to glycemic control, as measured by HbA1c [1, 2].

Four basic types of methods are used to measure HbA1c: immunoassay, ion-exchange high-performance liquid chromatography (HPLC), boronate affinity HPLC, and enzymatic assays. All the four methods are ineffective in assessment of glycemic control in patients homozygous for HbS, C or SC disease or any other conditions that reduce the life span of the erythrocytes. In HbAS, AC, AE, AD and F, the interference of results depend on the method of assay and the laboratories

should be aware of the limitations of their method with respect to these interferences, as it turned out in our case [3].

Other parameters of assessing glycemic control like frequent self monitoring of blood glucose (SMBG) and glycated albumin (fructosamine) may be used. In SMBG, cost of the glucometer strips, accuracy and repeated pricking are limiting factors. For fructosamine, the non-availability of the assay in many centers and the standardization of reporting is a problem. Fructosamine levels usually reflect the average glycemic control in the previous 2-3 weeks and the frequency of tests has to be decided based on that. With recent advances, continuous glucose monitoring system (CGMS) has been introduced where a catheter is inserted in the subcutaneous plane and is connected to a computerized glucose sensing apparatus. It aspirates micro-quantities of interstitial fluid at regular intervals and records the glucose values which may be analyzed later. The expected cost of the above system is a major limiting factor in a resource-constrained setting. Another test, though not approved by FDA, is 1,5 anhydroglucitol estimation whose concentration normally falls if blood glucose is above 180mg/dl. Hence, this is used to assess glycemic variability and reflects more of post-prandial control [4]. However, in a given situation like in our patient, these methods have to be resorted to once in a while to assess

the efficacy of the insulin regimen and diabetes control.

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#### REFERENCES

1. Diabetes Control and Complications Trial Research Group. The effect of intensive treatment of diabetes on the development and progression of long-term term

complications in insulin-dependent diabetes mellitus. *N Engl J Med.* 1993;329:977-86.

2. UK Prospective Diabetes Study (UKPDS) Group. Intensive blood-glucose control with sulphonylureas or insulin compared with conventional treatment and risk of complications in patients with type 2 diabetes (UKPDS 33). *Lancet.* 1998;352:837-53.
3. Little RR, Roberts WL. A Review of variant hemoglobins interfering with hemoglobin A1c measurement. *J Diabetes Sci Technol.* 2009;3:446-51.
4. Kim WJ, Park CY. 1,5-Anhydroglucitol in diabetes mellitus. *Endocrine.* 2013;43:33-40.

## Subacute Sclerosing Panencephalitis With Tics as First Symptom

A 10-year-old boy presented with complex tics involving shoulder and facial muscles since five months. Tics were insidious in onset, gradually progressive, changing in type and location, suppressed with effort. He suffered from measles at two years of age. Examination revealed stereotypic repetitive movements and bradykinesia. The initial provisional diagnosis was Tic disorder of childhood. After two weeks he developed spontaneous periodic generalised myoclonus followed by ataxia, progressive slurring of speech and decreased speech volume. Investigations showed normal complete hemogram, ESR, ASO titre, anti-nuclear factor, liver function test, serum electrolytes, copper, ceruloplasmin, parathyroid hormone and lactic acid levels. Nerve conduction studies, electromyography, axillary skin and muscle biopsy reports were normal. Computed tomography scan and magnetic resonance imaging (MRI) of the brain were normal. However, electroencephalography showed periodic generalised bursts of high-amplitude and slow wave complexes recurring at intervals of 6-8 seconds. Cerebrospinal fluid study revealed IgG measles 376.11 U/L (dilution 1:4) and serum IgG measles 329.09 U/L (dilution 1:404) by ELISA. Serum anti-measles antibody titre (5.22RFV) were elevated above normal range (>0.7RFV) by ELFA. Blood and CSF serology for Herpes simplex, Toxoplasma and Cytomegalovirus were all negative (both IgM and IgG). He was treated with Isoprinosine (100mg/kg/day) but therapy with interferon remained

unaffordable. Sodium valproate and clonazepam were added for control of myoclonus. Improvement in seizure control and tics were noted after six months of continuous follow up. Repeat MRI showed focal areas of hyperintensities in cortex and subcortical white matter of both frontal and adjacent high parietal region in T<sub>2</sub> weighted FSE and FLAIR image after two years of follow up.

Myoclonus is brief, involuntary twitching of a muscle or group of muscles, may be mistaken as tics and has been described with SSPE [1]. Tics are characterized by abrupt, repetitive movements, commonly preceded by premonitory sensation of an urge and can be suppressed with effort [2]. Tourette syndrome is most frequent cause of tics, others are insults to the brain; particularly the basal ganglia, infection, stroke, head trauma, certain toxins, drugs and various sporadic, genetic, neurodegenerative disorders [2]. Differential diagnosis of childhood cognitive deterioration and movement disorders like Wilsons disease, childhood systemic lupus erythematosus, hypoparathyroidism, Hallervorden Spatz disease and progressive myoclonic epilepsy were excluded due to lack of clinical, laboratory and imaging findings.

Previously reported atypical features of SSPE include isolated psychiatric manifestations, poorly controlled seizures, stroke like onset, hemiparesis, acute encephalopathy, cerebellar ataxia, visual disturbances, symptoms suggestive of intracranial space occupying lesion and parkinsonism like features [3-5]. While this association may be coincidental, but the possibility of tics as a result of insult to the brain due to SSPE should be kept in mind.

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