Hemichorea-hemiballismus associated with nonketotic hyperglycemia (HNKH) is clinically characterized by acute-onset involuntary movements involving one-half of the body. The classic triad includes nonketotic hyperglycemia, unilateral involuntary movements, and contralateral striatal abnormalities on neuroimaging. HNKH is commonly reported in elderly diabetic females, predominantly Asians. Pediatric HNKH is uncommon, with only 6 cases reported to date. We report a case of a 13-year-old girl who presented with HNKH as the initial manifestation of type 2 diabetes mellitus (DM).

A 13-year-old girl presented with a 3-week history of acute-onset, slow, nonrhythmic, variable-amplitude, involuntary movements involving left-side limbs; the lower limb was more affected, and movements were nonsuppressible and absent during sleep. There was no history of fever, sore throat, joint pain, increased thirst, polyuria, or significant weight loss. Other than involuntary movements, neurological examination did not reveal any focal deficits. Laboratory workup showed normal serum sodium and potassium levels, normal liver, renal and thyroid function tests, and normal anti-streptolysin O titer. Her random blood glucose level was 634 mg/dL, and her glycated hemoglobin (HbA1c) level was 12.4%. Serum osmolarity was 314.14 mOsm/L. Urinalysis for ketones was negative, and the C-peptide level was 2.8 ng/mL (normal = 0.81–3.85 ng/mL). Computed tomography (CT) of the brain revealed hyperdensity involving the right caudate, putamen, and globus pallidus, with subtle hyperdensity in the left basal ganglia (Supplementary Figure 1 in the online-only Data Supplement). Considering HNKH, insulin was started, along with symptomatic therapy including haloperidol 0.5 mg P.O. twice daily and clonazepam 0.5 mg P.O. once daily. Her blood glucose normalized over the next two weeks, and involuntary movements subsided at the one-month follow-up (Supplementary Video 1 in the online-only Data Supplement).

Hemichorea-hemiballismus may result from a vascular, autoimmune, infective, metabolic, or neurodegenerative pathology involving the basal ganglia. HNKH has been associated with poor glycemic control in diabetic patients. Nearly one-third (30.9%) of cases developed choreo-ballistic movements as an initial manifestation of DM. Including all reported adult HNKH cases in four major series, the mean age was 70.22 ± 10.85 years, and more than two-thirds (67.1%) of the cases were females. Including our case, HNKH has been reported as an initial manifestation of DM in five (71.4%) of the seven reported pediatric cases to date. The mean age of the pediatric HNKH patients was 13.71 ± 2.92 years (Table 1).

Our case was diagnosed 3 weeks after the onset of abnormal involuntary movements. HNKH is commonly misdiagnosed on the initial evaluation, with a delay of up to 6 months reported. The mean delay of 11.71 days (1–28 days) in the diagnosis of pediatric cases is lower than the that of 25.8 days (0–180 days).
days) reported in adults.1,2,5 Pediatric cases report slightly more deranged metabolic parameters, including serum glucose and HbA1c levels (Table 1). The mean serum glucose level was higher in pediatric cases (445.0 ± 212.6 mg/dL)1,4 than in adults (404.7 ± 199.8 mg/dL).1,2,5,6 The range of HbA1c levels reported in pediatric cases (12.0–17.3%)1,4 was higher than that in adults (9.7–14.4%).1,2,5,6 Our patient had a serum glucose level of 634 mg/dL, HbA1c of 12.4%, and serum osmolarity of 314.4 mOsm/L. In addition to hyperglycemia management, symptomatic therapy for involuntary movements is often used (80% of cases).1,6 Haloperidol, clonazepam and valproate are commonly used drugs.1,6 Including our case, five (71.4%) of the seven pediatric cases reported amelioration of involuntary movements within a month of onset.1,4 It remains unclear which clinical factors could predict the final clinical outcome. Neither baseline blood glucose nor clinical duration showed any clear association with the time to recovery or predicted neurologic sequelae. The involuntary movements in our case subsided with blood glucose normalization, along with the use of haloperidol and clonazepam. She had no involuntary movements at the one-month follow-up.

Our patient had hemichorea-hemiballismus predominantly affecting a lower limb. Unilateral involuntary movements were observed in 89.5% of adult cases, with the remaining cases developing bilateral choreo-ballistic movements.1,2,5,6 Both upper and lower limb involvement was observed in the majority of adult cases (93.3%), with additional involvement of the face reported in 20% of cases (Table 1).1,2,5,6 Occasional cases may have isolated upper limb involvement.1,5 In the pediatric age group, 85.7% of the cases reported involvement of both upper and lower limbs, with face involvement in 43% of the cases.5,6 Although two major series involving adult cases used the term “chorea” to describe involuntary movements in all reported cases,1,5 the other two series reported pure ballismus, choreo-ballistic movements and pure chorea, each in one-third of the cases.5,6 A single case presented with choreo-athetosis.5 Five (71.4%) of the seven pediatric cases manifested choreo-ballistic movements, with the other two having had pure chorea.5,6

Brain CT in our case revealed hyperdensity involving the right caudate, putamen, and globus pallidus, with subtle hyperdensity in the left basal ganglia. Well-characterized contralateral striatal hyperintensity on T1-weighted sequence in MRI and striatal hyperdensity on brain CT are commonly reported in HNKH, with bilateral abnormalities associated with bilateral chorea and/or ballismus and are rarely reported in hemichorea-hemiballismus.2,5 Neuroimaging findings may predate abnormal movements5 and have been reported even without involuntary movements in a patient with nonketotic hyperglycemia.7 Normal CT of the brain has been reported in three adult HNKH cases. Brain MRI showed putaminal hyperintensity in the first case, was normal in the second case and was unavailable in the third case.2,5 A normal MRI brain was reported in one

| Characteristics | Adult-onset cases (n = 95)1,2,5,6 | Pediatric-onset cases (n = 7)1,4 |
|-----------------|---------------------------------|-------------------------------|
| Demographic data |                                 |                               |
| Age (yr), mean ± SD (range) | 70.22 ± 10.85 (22–92) | 13.71 ± 2.92 (8–17)          |
| Females (%) | 67.1 | 57.1          |
| Asians (%) | 63.2 | 57.1          |
| Delay in diagnosis (day), mean (range) | 25.85 (0–180) | 11.71 (1–28) |
| Clinical presentation |                                 |                               |
| Unilateral involuntary movements (%) | 89.5 | 100           |
| Both upper and lower limbs involved (%) | 93.3 | 85.7          |
| Face involved along with upper and lower limbs (%) | 20 | 42.8          |
| Laboratory investigations |                                 |                               |
| Serum glucose (mg/dL), mean ± SD | 404.7 ± 199.8 | 445.0 ± 212.6 |
| HbA1c range (%) | 9.7–14.4 | 12.0–17.3 |
| Serum osmolarity range (mOsm/L) | 304.8–313.7 | 284–318 |
| Findings on brain MRI or CT |                                 |                               |
| Cases with putamen involvement (%) | 100 | 100           |
| Cases with caudate involvement (%) | 50 | 100           |
| Treatment |                                 |                               |
| Use of symptomatic therapy, yes (%) | 80 | 85.7          |
| Most commonly used symptomatic therapy | Haloperidol | Haloperidol |

HbA1c: glycosylated hemoglobin.
of the pediatric cases. While the putamen is universally affected, the caudate and/or globus pallidus may be involved in up to 50% of adult cases, but all pediatric cases had neuroimaging abnormalities (Table 1). Although striatal lesions may persist in a few cases, radiological resolution is commonly observed following clinical improvement. Since most HNKH cases show clinical and radiological recovery, incomplete basal ganglionic dysfunction seems to be the likely pathology. Although the pathophysiology of HNKH is not clear, several factors, including metabolic dysfunction, vascular insufficiency, petechial hemorrhages, and hyperviscosity, are believed to contribute.

HNKH is an uncommon choreo-ballistic movement disorder that is commonly reported in elderly females. It may rarely occur in the pediatric age group, even as an initial manifestation of DM. Early recognition and prompt management of serum glucose, with symptomatic therapy for involuntary movements, often result in complete clinical improvement in the majority of cases.

Ethics Statement
Written informed consent was obtained from the patient.

Supplementary Video Legends
Video 1. The first part of the video shows the pretreatment status with hemichorea-hemiballism involving the left upper and lower limbs. The second part of the video, recorded after normalization of serum glucose and use of symptomatic therapy, shows no involuntary limb movements during follow-up on day 30.

Supplementary Materials
The online-only Data Supplement is available with this article at https://doi.org/10.14802/jmd.20128.

Conflicts of Interest
The authors have no financial conflicts of interest.

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Supplementary Figure 1. Brain CT in our case, showing diffuse hyperdensity of the right caudate and lentiform nuclei (A, B) and subtle hypodensity involving the left caudate and lentiform nuclei (B).