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Slipped capital femoral epiphysis in a patient with dravet syndrome: A case report and literature review

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Abstract: In situ fixation of SCFE is the widely accepted treatment for both stable and unstable SCFE. Prophylactic fixation of the contralateral hip remains controversial. There is still no agreement about the removal of the screws in the absence of implant-related symptoms.

The case

- A case of a slipped capital femoral epiphysis SCFE in an adolescent with Dravet Syndrome after a seizure attack.
- To the best of our knowledge, this is the first report worldwide about the presentation of SCFE in Dravet syndrome.
- Dravet syndrome is severe myoclonic epilepsy in infancy.
- Cases of SCFE after epileptic fit need more studies and long-term follow-up to understand the pathophysiology and consequences of treatment.

Lessons learnt

- This rare case needs more follow-up till adulthood.
- Possible complications and further management are challenging culprits.
- Children presented with SCFE after epileptic fit, withdraw special attention. However, there is a lack of knowledge in the literature about their natural history & long-term follow-up.

Keywords: slipped capital femoral epiphysis, dravet syndrome, epilepsy, seizure

Introduction

Dravet Syndrome (DS) is clinically characterized as early as childhood by pleomorphic seizures and demonstrates neurodevelopmental delay, and cognitive and motor disability^[1]. Other manifestations like intellectual disability, ataxia and crouching gait, could be seen with ageing^[2]. Treatment modalities include anti-epileptic drugs and ketogenic diet therapy in addition to surgical options such as deep brain and vagal nerve stimulation^[3].

Slipped capital femoral epiphysis (SCFE) is a common cause of non-traumatic painful hip in adolescents. Long-term effects of SCFE could be irreversible and may lead to early onset impairment and the need for early hip reconstruction surgery^[4]. Obesity is considered the most important risk factor for SCFE. Others include male sex, periods of rapid growth, and prior radiation therapy. Regarding onset, the average age in females is 11.2 years, and in males is 12.0 years. Bilateral cases are approximately 25% (Range: 8-50%)^[5]. The possible complications are avascular necrosis of the femoral head (AVN) and femoro-acetabular impingement (FAI). Others include early onset hip osteoarthritis, and implant-related complications^[6]. In situ fixation of the upper femoral physis with a partially threaded cannulated screw is considered the most efficient stabilization modality for both types of SCFE, stable and unstable^[6].

Case presentation

A 14-year-old boy presented with an unstable type of SCFE in the left hip. He is a known case of Dravet syndrome, Genetic study had reported SCN2A gene mutation, with uncontrolled recurrent attacks of seizures for which he has been receiving sodium valproate and levetiracetam. He had complained of left hip pain and lost gait function over two weeks after a febrile seizure. Upon examination, He was holding bilateral hip and knee joints in flexion position and passive extension of the left hip was painful. X-ray bilateral hip joints had confirmed slipped capital femoral epiphysis of the left hip (Figure 1). Parents were counselled for surgical intervention. Informed and written consent was taken. He underwent in-situ fixation of the left hip with a partially threaded cancellous screw 6.5 mm (Stainless steel-Synthes) (Figure 2).



Fig 1: Radiographs of pelvis & left hip showing Slipped Capital Femoral Epiphysis in the left hip at first presentation in Accident & Emergency Department.

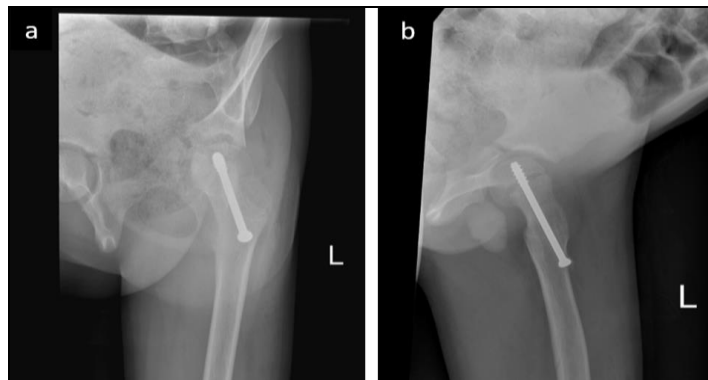


Fig 2: Radiographs of left hip anteroposterior (a) and lateral (b) views immediate post-surgery with in-situ fixation by partially threaded cancellous screw.

He had been discharged home with advice for non-weight-bearing mobilization. At a six-week post-operative follow-up, he started partially assisted weight-bearing. At 12-week

postoperative, he started full weight bearing without support (Figure 3).

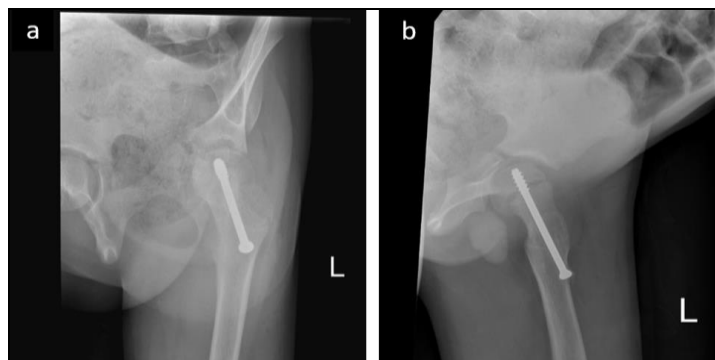


Fig 3: Radiographs of left hip anteroposterior (a) and lateral (b) views at 12-week follow-up.

One year postoperative, he can walk independently – crouch gait – near the normal range of motions in hip joints in

comparison to the contralateral side (right hip) till the last follow-up at 36-week (Figure 4).

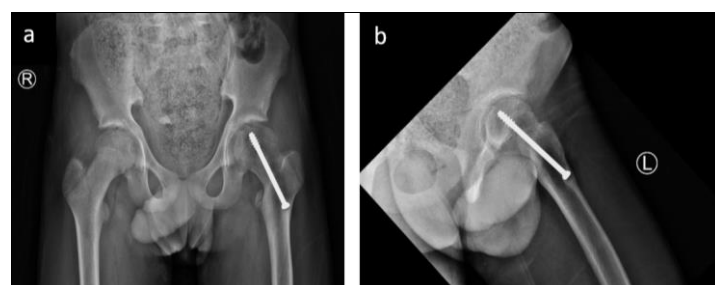


Fig 4: Radiographs of left hip anteroposterior (a) and lateral (b) views after 36 weeks follow-up post-surgery.

Fortunately, during that time, the contralateral slippage had not been encountered despite continuation of uncontrolled fits – an average of 2-3 fits/month – with active medical treatment.

Discussion

Unilateral SCFE in an adolescent with Dravet syndrome

Table 1: Published case reports of SCFE after an epileptic fit.

	No. of cases	Age	side	Treatment	Contralateral Slippage
Patterson <i>et al.</i> [7]	1	12-year	Right	Percutaneous pinning	After 8 months
Yilmaz <i>et al.</i> [8]	1	12-year	Bilateral	In situ fixation	Simultaneous
Nhamoucha <i>et al.</i> [9]	1	15-year	Right	In situ fixation	Not reported
Kyriakos P <i>et al.</i> [10]	1	10-month	Left	Closed traction and a hip spica cast	Not reported

Patterson *et al.* reported a case of a 12-year-old with right side SCFE after an epileptic seizure who developed contralateral slippage after 8 months from surgery [7]. Yilmaz *et al.* had reported a case with the same age but presented with bilateral SCFE following epileptic fit [8]. A case review had published by Nhamoucha *et al.* on a 15-year-old adolescent with right side SCFE after an epileptic seizure who did not report contralateral slip [9]. A left SCFE had occurred in a 10-month-old infant post-seizure attack as described by Kyriakos P *et al.* which was uncommon for this age group [10].

The percentage of contralateral slip in patients with primary unilateral slip varies from 8% to 36% till skeletal maturity [11]. Prophylactic fixation of unaffected hip in a patient with a unilateral SCFE remains controversial. The argument against routine prophylactic fixation is the potential risk for surgical complications [12]. Despite multiple risk factors that have been studied, including young age, obesity, endocrine disorders, and increased posterior sloping angle, there is currently no precise and universally accepted method of predicting contralateral slip; this important subject demands further research [13].

After the closure of the physis, routine implant removal following termination of growth around puberty is controversial keeping hardware in place may result in fractures in young adults secondary to stress risers and difficulty with future arthroplasty [14]. On the other hand, implant removal requires a second planned operation, with increased surgical exposure, implant removal failure and risk of fractures at the time and after implant removal [15]. It is recommended that SCFE stabilization screws should be removed only if they have developed secondary symptoms like tendinitis of the iliotibial band, bursitis of the greater trochanter or screw loosening or migration. To date, there are no clear indications to remove or not an asymptomatic metal and the surgeon should weigh the risk-benefit ratio [4].

Conflict of interest: Nil

Source of support: Nil

Consent

The authors confirm that the informed consent of the patient is taken for the publication of this case report

Abbreviations: Dravet Syndrome (DS), Slipped capital femoral epiphysis (SCFE). Avascular necrosis of the femoral head (AVN) and Femoro-acetabular impingement (FAI).

Following an epileptic seizure is a unique presentation. On review of the literature, few case reports have been published with SCFE after epileptic fit (Table 1). The exact pathophysiology of SCFE after an epileptic fit is still unclear but forces transmitted during the seizure across the weakened physis may be the cause.

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