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Recurrent hip dislocations in a child with congenital insensitivity to pain with anhidrosis (CIPA) syndrome: a case report.

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ABSTRACT

congenital insensitivity to pain with anhidrosis (CIPA) syndrome is a rare autosomal recessive disease, with children affected presenting with frequent musculoskeletal manifestations.

In this case report we present the case of a 4 years old female child with recurrent hip dislocation, which is managed conservatively with closed reduction and bracing. The child continues to have spontaneous dislocations that can be reduced at home.This case discuss the challenges dealing with dislocated hips and potential sequela of Charcot joint.

1. INTRODUCTION

CIPA syndrome is a rare autosomal recessive disorder (1 in 125,000,00) (1). A mutation in NTRK1 gene was identified (2), the hallmark of the disease is loss of sensation and anhidrosis. They usually present with unexplained fever at birth and delayed developmental milestones, tongue and toes ulcerations(3).

Musculoskeletal manifestations are an essential part of the disease process, as the lack of protective sensation will lead to fractures, ulcers, dislocations and charcot joint(3,4).

We present a 4 years old female with CIPA syndrome presented with recurrent hip dislocations.

2. CASE PRESENTATION

Our patient is the first child, female, product of normal delivery, full term. She was normal at birth with no concerns by family or doctors. Following discharge next day after birth the parents noticed persistent fever around 40 degrees, she was brought back to hospital and full workup for infection was done and came back negative.

In the first year of her life parents noticed delayed development, like inability to sit and generalised hypotonia, a pediatric neurologist was able to suspect diagnosis and genetic testing was done to confirm diagnosis. The patient was found to have homozygous mutation of NTRK1 gene.

The patient started crawling around her first birthday and walking at 18 months of age, since age one she suffered multiple skeletal injuries including: foot ulcers, foot metatarsal fracture, and multiple lower sprains.

At age 34 months she presented with the first time with left hip dislocation, the parents reported a minor trauma at home, as she was on the sofa and fell down, then mother noted her left leg is shorter and brought her to the emergency department (ED). Upon examination, she had good vascularity; the leg was shortened and internally rotated. Plain imaging performed and confirmed left hip dislocation (Figure 1).

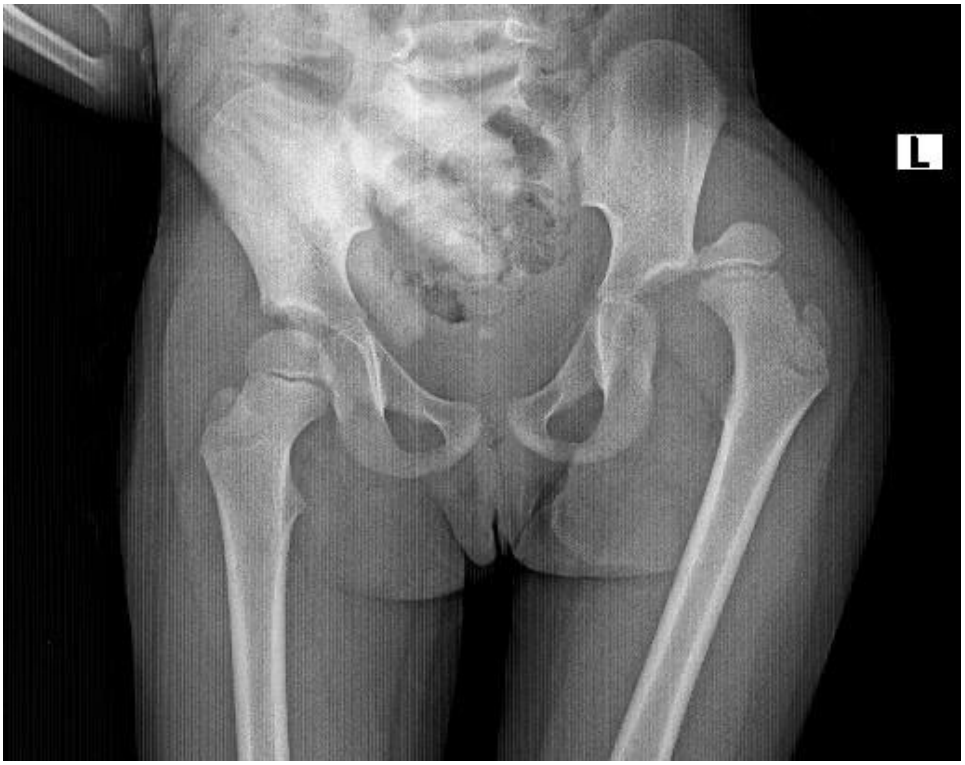

Figure 1

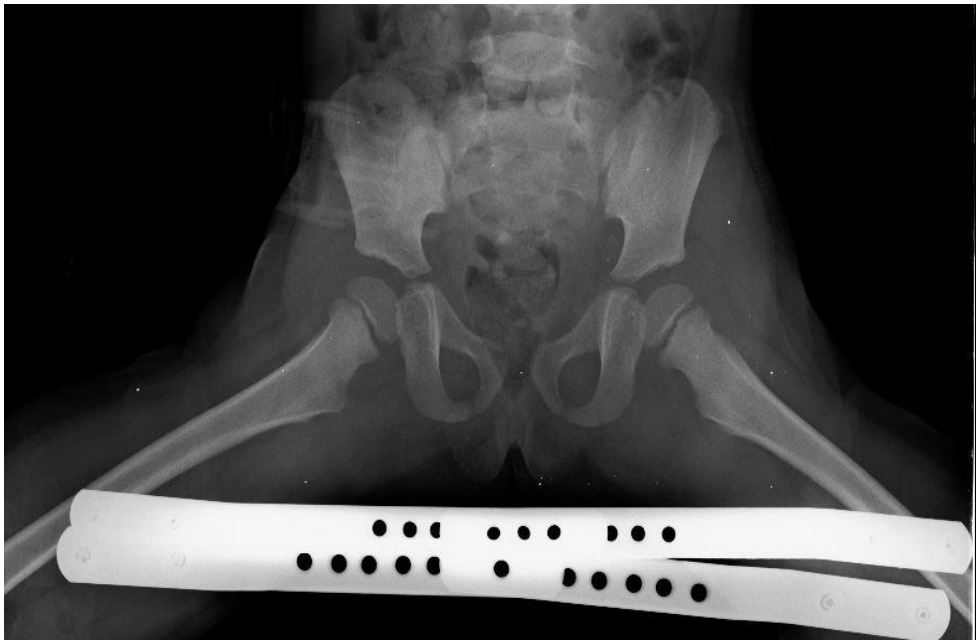

Figure 2

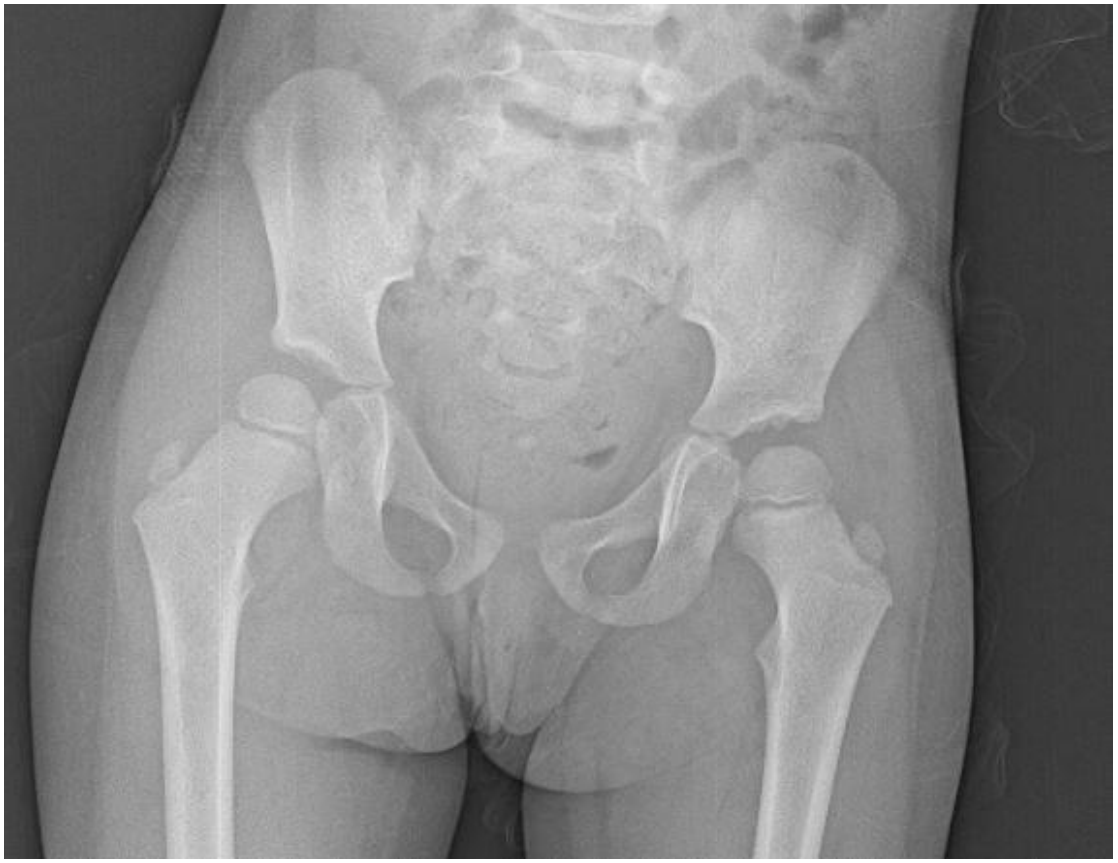

Figure 3

Since she doesn't have any pain the hip was successfully reduced in ED, and the leg placed in an abduction hip brace for 6 weeks following confirming reduction with plain radiograph (Figure 2). At the end of 6 weeks another radiograph was obtained and the hip can be seen reduced in socket (Figure 3).

The patient presented again 7 months later with dislocation on the same side, as she was running at home. Reduction was achieved and a brace was used as the first time. No significant changes on radiographs were noted. The patient was seen 6 months following the last incident, now parents report she had dislocations on the same side every 2 months on average, and they didn't need to come to hospital as they were able to reduce at home.

Other issues she is having due to her condition include: left big toe wound failure to heal for over 3 months, she suffered gluteal abscess on the right buttock drained a year ago and followed by full recovery

3. DISCUSSION

The case we presented explain the complex nature of the disease and the difficulty managing these patients by their parents and medical professionals, as multidisciplinary approach should be used. Parent education about the condition, how to manage and what to expect is of crucial importance.

Musculoskeletal complications of CIPA syndrome are frequent and can be challenging to manage. A wide variety of complications can occur including: fractures, dislocations(5), ulcers, charcot joint and infections (6,7,8).

Most of the cases with hip dislocations were managed conservatively, with reduction and bracing in most cases (6). Surgery should be avoided if possible in these case, as wound healing and lack of protective sensation increase the possibility of complications (4).

4. CONCLUSION

Hip dislocations in CIPA syndrome patients is not uncommon among this small group of patients; most cases tend to have recurrent dislocations. Charcot joint is the likely sequela.

Family education remains the most important factor to limit recurrence and other injuries in these patients. As reducing the

number of dislocations could avoid the potential joint damage and need for surgical interventions, which has high risk of complications.

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