Introduction:
Complex Regional Pain Syndrome (CRPS) is rare in the pediatric population with an uncharacterized etiology and risk factors. Over the course of one year, four patients presented to our pediatric chronic pain clinic with CRPS and were found to have significant joint hypermobility with frequent joint trauma. More significantly, these patients' symptoms were compatible with the diagnosis of Ehler-Danlos Syndrome, Type 3. Of note, the four patients represent 50% of patients that presented with CRPS during that time period. This is a novel case series because Ehlers-Danlos Syndrome (EDS) has not previously been described as a predisposing factor for the development of complex regional pain syndrome in children.

Description:
Patient #1 is a 13 year-old female who presented with CRPS of the left lower extremity. She has a history of frequent sprains of the bilateral knees and right ankle and severe chronic pain of the left knee after a sprain. Upon further examination, she was determined to have many signs of EDS including hypermobility of multiple joints, striae of the abdoman and upper extremities, and a Beighton score of 5/9. Genetic testing was completed and she was determined to have EDS, Type 3.

Patient #2 is a 16 year-old female who presented to the pain clinic with CRPS of the left lower extremity. She has had severe chronic pain since Spring 2014 after a minor ankle sprain. She has a history of hypermobility of multiple joints, frequent ankle sprains, and "being double-jointed." Her Beighton score was 7/9 in clinic. She underwent genetic evaluation and was determined to have EDS, Type 3.

Patient #3 is a 16 year-old female who presented to chronic pain clinic with a history of CRPS of the right upper extremity and left lower extremity since 2012. Her past medical history was significant for frequent headaches, abdominal pain, and autoimmune thyroiditis.

Patient #4 is a 7 year old female who presented with a diagnosis of CRPS of the left lower extremity. At the initial clinic visit, the family described a long history of knee and ankle pain. She had hypermobility in bilateral ankles, wrists, and fingers with a Beighton score of 7/9. Striae were noted on examination. Family history was positive for EDS in both her mother and maternal grandfather. She underwent genetic evaluation and was determined to have EDS, Type 3.

Discussion:

- Ehlers-Danlos Syndrome (EDS) is a connective tissue disorder with several variants.
- EDS Type 3 can involve skin hyperextensibility, joint hypermobility, headaches, abdominal pain, widespread musculoskeletal pain, joint dislocations, joint subluxations, frequent sprains, easy bruising, and temporary paresthesias from nerve compression.
- Symptoms compatible with EDS Type 3 were commonly found in patients presenting to the Children’s Pediatric Pain Clinic with CRPS.
- EDS/hypermobility may have an association with the development of CRPS in the pediatric population. Future studies are warranted to ascertain whether EDS and/or hypermobility are risk factors for the development of CRPS.
- Additionally, pediatric patients presenting with CRPS should be screened for joint hypermobility and EDS Type 3.

References: