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StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2018 Jan-.

Klumpke Palsy

Authors

Justin Merryman; Matthew Varacallo[1].

Affilations

[1] Department of Orthopaedic Surgery, University of Kentucky School of Medicine

Last Update: October 27, 2018.

Introduction

Klumpke palsy, named after Augusta Dejerine-Klumpke, is a neuropathy involving the lower brachial plexus.[1] In contrast, the more common Erb–Duchenne palsy involves the more cephalic portion of the brachial plexus C5 to C6.[2] The brachial plexus is a bundle of individual nerves that exit between the anterior and middle scalene muscles in the anterior lateral and basal portion of the neck. Although, the most common anatomical presentation of the brachial plexus is between the anterior and middle scalene there are variations with the most common being penetration of the anterior scalene.[3] The main mechanism of injury to the lower brachial plexus is hyper-abduction traction and depending on the intensity, will lead to signs and symptoms consistent with a neurological insult.[4]

Etiology

The most common etiology resulting in Klumpke palsy is a hyper-abduction trauma to the arm that has enough intensity to traction the lower brachial plexus. Trauma during birth can cause brachial plexus injuries, but again hyper-abduction and traction forces to the upper extremity are usually present. A compression injury has similar signs and symptoms, for example, an apical lung tumor gets to the size it presses on the cervical sympathetic ganglia and C8 to T1 nerve root if there is a sympathetic involvement Horner syndrome may present.[5] Approximately 50% of all brachial plexus injuries happen between the age of 19 to 34.[6]

Epidemiology

Of 4538 multi-trauma patients, Midha observed brachial plexus injuries in 1.2%; 62% were supraclavicular, and 38% were infraclavicular C8 to T1. A motor vehicle accident caused the most injuries. Supraclavicular accidents have higher grades of neurological damage based on Sunderland classification, while infraclavicular injuries resulted in neurapraxia more often.[7] Another study found a similar prevalence amongst 203 patients 90% were supraclavicular, and 10% infraclavicular, interesting, 47% patients had no prior pain while 28% did.[8]

Pathophysiology

The pathophysiology of this condition is one of neurological insult. For there to be signs and symptoms, first there must be enough injury to the nerves. Seddon and Sunderland developed one way to understand this nerve compromise by examining the injury micro and macroscopically. Nerve compromise has 3 main categories; neurapraxia, axonotmesis, and neurotmesis. During the neurapraxia state, there is no macroscopic damage, but there is functional damage, most are self-resolving with physiotherapy. The other 2 categories, axonotmesis and neurotmesis, are more serious cases as there is macroscopic damage with neurotmesis having the less ideal prognosis usually resulting in a non-functional neuroma. The most common findings at all stages of neurological degeneration would be dermatome and myotome abnormalities specific to nerve roots.[5]

History and Physical

The history presented by the patient usually depicts a long axis hyper-abduction traction injury with high amplitude and velocity.[9] The typical patient presentation is a decrease of sensation along the medial aspect of the distal upper extremity along the C8 and T1 dermatome. The patient might also present myotome findings that can range from decreasing muscular strength to muscular atrophy and positional deformity. For example, if the neurological damage has led to muscular atrophy and tightening the patient may present with a "claw hand." This deformity presents as finger and wrist flexion. The patient may also describe the severe pain that starts at the neck and travels down the medial portion of the arm. One other sign of a lower brachial plexus injury is Horner syndrome, because of its approximation to the T1 nerve root it may damage the cephalic sympathetic chain. If this happens, the patient will develop ipsilateral ptosis, anhidrosis, and miosis.[5][10]

Evaluation

Evaluation of this injury would start out with a thorough patient history and physical examination. During the physical examination, the practitioner would assess the involved upper extremity if neurological findings are present diagnostic imaging would follow. Cervical and thoracic radiographs along with computed tomography CT or magnetic resonance MR are all valuable tools. Conventional radiographs will at most rule out and neurological compromise secondary to hard tissue, for example, fracture impeding the nerve roots. With nontraumatic cases, conventional radiographs may also rule out any space-occupying lesions in the superior mediastinum and basal portion of the neck. CT and MR evaluation are more precise when considering a neurological insult. CT myelography is a more specific and sensitive evaluation. MR contributes less radiation, but it also has less sensitivity and specificity compared to CT myelography. Other tests could include a histamine injection and nerve conduction velocity NCV. They use the histamine test to approximate the neurological injury proximal or distal to the dorsal root ganglion if the injection if histamine causes an intense dermatomal "flare" reaction the insult is proximal to the dorsal root ganglion. An NCV test measures how fast neurological impulses travel and can locate the injury.[5]

Treatment / Management

Because of its location, Klumpke palsy can also affect the axillary artery; this can be from both a hyper-abduction stretch injury, but also a crush injury to the local area. If there is a severe vascular compromise, the patient will undergo emergency surgery. Fractures correlate with this condition and treatment typically involves surgery. With injuries that are closed, meaning they do not have an obvious break through the skin or are vascular compromised there is a conservative option. Spontaneous recovery from nerve palsy is possible, but it is important to note that most C8-T1 nerve palsies do not recover without intervention. Conservative treatment including exercising of supportive muscles and stretching affected muscles helping to maintain the range of motion and aid in a full recovery. The nerves will heal, but most are mild cases. In 3 to 6 months, if there is no improvement, surgery will take place. Surgical correction mostly involves nerve grafting, but in cases of injuries proximal to the dorsal root ganglion, neurotization is the recommended procedure.[11]

Differential Diagnosis

Similar signs to Klumpke palsy is Erb's palsy; this injury affects the upper brachial plexus which will usually result in dermatome and myotome finds along the C5-C6 path, for example, someone with Erb's palsy usually has the "waiter tip" presentation, fixed pronation of the forearm with an outward facing palm.[2] Another similar condition is distal nerve entrapment of the ulnar nerve at either the medial epicondyle of Guyon's tunnel. It is referred to as ulnar nerve entrapment and can produce similar neurological finds as the more proximal Klumpke palsy. One big difference is that there will be no involvement of innervation proximal to the lesion, for example, you will not see pectoralis major involvement with true ulnar nerve entrapment.[12] Thoracic outlet syndrome (TOS) can also show similar symptoms, typically TOS is a compression injury to the brachial plexus from a rudimentary rib, first rib, or the clavicle on the ipsilateral side, this could be post-traumatic, postural driven, and or genetic. In contrast to Klumpke, TOS will affect more than just the C8-T1 nerve roots, but like Klumpke it also can affect the axillo-subclavian artery.[13] Below is a list of the most common differential diagnosis to Klumpke palsy.

- Distal ulnar nerve entrapment
- Thoracic outlet syndrome
- Apical lung tumor
- Neurofibroma
- Disc herniation
- Shoulder impingement
- Clavicular or vertebral fracture
- Others

Prognosis

A 2006 study found a better prognosis in children with a brachial plexus lesion compared to adults undergoing microsurgery. An explanation for this might have to do with a shorter distance, a higher threshold for regeneration, and an increased cognitive adaptability.[14] Another article showed similar findings that microsurgery helped to restore hand function in 75% of the cases in 8 years.[15] Spontaneous recovery is possible with less intense injuries. Zuckerman and colleagues found that in similar groups one undergoing surgery and another past the point of surgery had a similar functional outcome in 2 years.[16] Most professions will agree a better prognosis for more severe injuries with a shorter time to the onset of surgery.

Postoperative and Rehabilitation Care

With brachial plexus birth palsy, if there is no fracture or dislocation present, exercise focusing on stretching and strengthen restricted muscles will help in functional recovery.[17] Goals of rehabilitation include; increasing range of motion, preventing muscular atrophy, and pain control.[6] Stretching of the tissue will aid in the prevention of muscular atrophy, the repetitive stretch has shown positive results with limiting atrophy in rats.[18] Other than stretching, electrical stimulation might also provide some therapeutic benefit. In a rat model, electrical stimulation has positive results with the denervated soleus muscle, but they observed limitations.[19]

Enhancing Healthcare Team Outcomes

Brachial plexus injuries are often difficult to manage as a portion of them recover spontaneously. It is important to develop strict guidelines that will get patients to surgery faster. The professional who is monitoring the patient must track the progression or regression of the neurological symptoms. In more severe cases, if the time to surgical correction is less than 6 months, there is a greater prognosis for muscular recovery. Exactly 89.7% of the patients had grade 3 muscular recovery when they underwent surgery at 3 months, and at 12 months, the number was only 35.7%. [6][20] (Level I)

Questions

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Bookshelf ID: NBK531500 PMID: 30285395