

## Vaughn-Jackson Syndrome

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### Abstract

*Vaughn-Jackson Syndrome is a specific pathophysiologic process that leads to extensor tendon rupture in a rheumatoid wrist. While uncommon due to the introduction of disease-modifying anti-rheumatic drugs, early identification and referral to an orthopaedic surgeon is vital to patient outcomes.*

**Keywords:** arthritis, rheumatoid, Tendons/injuries, Tendons/surgery, Tenosynovitis, Wrist Joint/abnormalities

### BACKGROUND

Rheumatoid arthritis is a chronic progressive systemic inflammatory disease. It is the most common inflammatory arthritis, with >90% of patient's having wrist symptoms 10 years after diagnosis.<sup>1,2</sup> The introduction of disease-modifying anti-rheumatic drugs have drastically improved patient's lives and lowered the number of patient's requiring orthopaedic intervention.<sup>2</sup> The 2015 American College of Rheumatology Guideline for the Treatment of Rheumatoid Arthritis details the medical treatment algorithm for rheumatoid arthritis.<sup>3</sup> This includes a combination of DMARD's and corticosteroids. One such problem highly associated with a rheumatoid wrist is Vaughn-Jackson syndrome. Vaughn-Jackson syndrome was originally defined in 1948 by Dr. OJ Vaughan-Jackson who described a unique set of two patients with whom had extensor tendon rupture due to attrition at an arthritic distal radio-ulnar joint.<sup>4</sup> Before this time, extensor tendon ruptures of the extensor pollicis longus as it traverses around Lister's tubercle of the distal radius from long standing tenosynovitis had been well defined. However, Vaughn-Jackson syndrome is unique in that treatment is directed at the pathologic distal radio-ulnar joint

and ulnar head in addition to repair of the extensor tendons.

### PATHOPHYSIOLOGY

Extensor tendon rupture in the rheumatoid wrist can be due to multiple etiologies. These include attrition of the tendon repeatedly rubbing against an osteophyte bony prominence, poor blood flow into the tendon possibly due to pressure thereby weakening the tendon, or infiltration of the tendons by inflammatory synovium.<sup>4-7</sup> Attrition can happen at many areas of the rheumatoid wrist, including flexor pollicis longus rupture due to bony spurs/osteophytes in the carpal tunnel.<sup>8</sup> Dr. Vaughn-Jackson described a pathologic process where an arthritic roughening of the articular margin of the distal end of the ulna erodes through the joint capsule of the wrist, directly contacting the extensor tendon to the fifth digit leading to tendon rupture.<sup>4</sup> If treatment is delayed, the neighboring intact extensor tendons naturally migrate ulnar-ward towards the defect left by the ruptured the tendon, and the patient will subsequently develop extensor tendon rupture of the fourth, then third, and finally second digit.<sup>9</sup> Therefore it is not unusual to have a patient first rupture the extensor tendon to the fifth

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digit, then progressively rupture extensor tendons of the remaining digits in a radial direction prior to presentation.<sup>9</sup> Risk factors correlating significantly with spontaneous extensor tendon rupture include disease duration longer than 8 years, persistent tenosynovitis longer than 1 year duration, and Larsen grade greater than 4 (severe erosions of the bone and joint). Dislocation of the distal end of the ulna, carpal collapse and the scallop sign on X-ray (erosion of the distal ulna into the radius at the distal radio-ulnar joint) contribute to a higher spontaneous extensor tendon rupture rate among rheumatoid arthritis patients.<sup>6,10</sup>

### DIAGNOSIS

Extensor tendon ruptures usually present as a

painless and sudden loss of finger extension. The rate of extensor tendon rupture in rheumatoid arthritis has been found to be approximately 1.6%.<sup>11</sup> In the case of Vaughn-Jackson syndrome, this sudden loss of finger extension usually begins at the fifth digit and proceeds radial-ward towards the second digit in a progressive fashion. Therefore, some individuals may present with multiple extensor tendon ruptures and not only an isolated rupture of the fifth extensor tendon.<sup>9</sup> On physical exam, there is dorsal wrist swelling, prominence of the ulnar head dorsally, loss of tenodesis effect during passive wrist motion, loss of palpable tendon tension with attempted active extension, and inability to actively hold the metacarpophalangeal joint in extension after passive extension of the joint.<sup>4,12</sup> (Figure 1)



**Figure1.** Patient with tendon rupture of the extensor tendons to the third, fourth and fifth fingers. Note the prominent ulnar head.

Other causes of loss of finger extension must be ruled out, including extensor tendon subluxation, posterior interosseous nerve compression, and ulnar clawing.<sup>12,13</sup> Extensor tendon subluxation is due to chronic inflammation of the metacarpal phalangeal joint causing the tendon to subluxate, usually ulnarly, leading to a flexed ulnarly deviated finger. (Figure 2) The patient is unable to actively extend the flexed finger; however, when the finger is passively extended by the examiner, the extensor tendon centralizes and the patient is able to actively maintain the digit in full extension. The inability to extend the digits associated with posterior interosseous nerve compression results

from rheumatoid synovitis at the elbow.<sup>13</sup> In contrast to extensor tendon rupture, the patient will also have weakness to extensor pollicis longus and abductor pollicis longus as well. (Figure 3) In a low ulnar nerve palsy at the wrist, the hand is held in an “ulnar claw” due to intact pull of extensor and flexor tendons, but loss of the ulnar nerve innervated intrinsic muscles of the hand. This causes hyperextension at the metacarpophalangeal joint and finger flexion at the proximal and distal interphalangeal joints. (Figure 4) Additionally, there is also weakness to pinch and grip, as all the ulnar nerve innervated intrinsic hand muscles are effected.<sup>14</sup>

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X-rays are required to delineate the degree of ulnar head and distal radio-ulnar joint arthritis. (Figure 5) To help make the diagnosis of Vaugh-Jackson syndrome, advanced imaging can be performed. CT is rarely required but can be used to define the bony anatomy of the ulnar head and distal radio-ulnar joint.<sup>15</sup> MRI is recommended if there is a question

of continuity of the extensor tendons.<sup>16</sup> Sometimes a final diagnosis cannot be made until surgical intervention.<sup>9</sup> In general, a multi-disciplinary team consisting of a rheumatologist, orthopaedic surgeon and occupational therapist are required to ensure these patients are accurately diagnosed and cared for.



**Figure2.** *Extensor tendon subluxation with metacarpal phalangeal joint swelling and ulnar deviation of the fingers.*



**Figure3.** *Posterior interosseous nerve palsy with loss of extension to the wrist, thumb, and fingers.*



**Figure4.** *Ulnar clawing with hyperextension of the metacarpal phalangeal joints of the fourth and fifth fingers with flexion of the proximal and distal interphalangeal joints.*



**Figure5.** *X-ray of a rheumatoid wrist with extensive erosions and arthritic changes, including the ulnar head.*

### SURGICAL TREATMENT

In order to restore a patient's function with Vaughn-Jackson syndrome, surgery is aimed at addressing the offending structures (ulnar head arthritis and synovitis) as well as repairing the extensor tendon ruptures. To resolve the primary pathophysiology of the bony erosion and osteophytes of the ulna causing an attritional rupture of the extensor tendons, a Darrach procedure is performed where the distal ulna is resected and capsule repaired.<sup>7,12,17</sup> Alternatively, an ulnar head arthroplasty (replacement) has demonstrated similar outcomes with its own unique advantages.<sup>18</sup> In treating the ruptured tendons, tenosynovectomy is combined with a side-to-side transfer of the ruptured distal tendon stump to a nearby functioning extensor tendon.<sup>7,12</sup> In cases of multiple extensor tendon ruptures without the ability to transfer to a nearby extensor tendon, a flexor tendon can be transferred through or around the forearm to the ruptured ends of the extensor tendons.<sup>7,12</sup> Extensive tenosynovectomy of the remaining extensor tendons is performed as recurrent tenosynovitis after synovectomy is rare (0-7%).<sup>6,12</sup> Certified hand therapy is an essential component of postoperative treatment in the patient's recovery of function.<sup>2,19</sup>

### CONCLUSION

In conclusion, Vaughn-Jackson syndrome is a specific pathophysiologic mechanism by which a diseased distal radio-ulnar joint leads to subluxation of an arthritic ulnar head dorsally, eroding away at the extensor tendons of the hand leading to subsequent tendon rupture and loss of finger extension. It can usually be diagnosed with history and physical exam but confirmed with plain X-ray and advanced imaging in questionable cases. The treatment is surgical with a combination of tenosynovectomy, tendon transfers, and distal ulna resection versus arthroplasty. Early referral to an orthopaedic surgeon is paramount before additional extensor tendons rupture, and a multi-disciplinary approach to these patient's care is essential to good outcomes.

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