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SUPPLEMENT ARTICLE

Pathology of the Elbow in Haemophilia

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ABSTRACT

In this article, we have reviewed the most important modern conceptual developments for the treatment of the elbow in persons with haemophilia. Despite modern preventive strategies, subclinical bleeding can still occur, underscoring the need for constant vigilance. The elbow is particularly susceptible to synovitis due to its extensive synovial tissue and multiplanar movement, which increase exposure to recurrent bleeding episodes. Iron deposition in the joint initiates a complex inflammatory and apoptotic cascade that ultimately leads to changes in the synovial membrane. Point-of-care ultrasound has become an essential tool for detecting subclinical synovitis, but ensuring diagnostic accuracy remains critical. Comprehensive treatment also requires a highly specialised musculoskeletal expert with access to the full range of therapeutic options. When conservative measures fail, rapid deactivation of the synovial tissue is imperative, and synovectomy stands out as a simple but highly effective intervention. In addition, advances in haematological care now allow the entire range of orthopaedic reconstructive procedures on the elbow to be performed safely in patients with haemophilia. In this evolving context, the role of a coordinated and specialised multidisciplinary team has become more important than ever, as it ensures the full integration of medical, rehabilitative and surgical approaches to optimise long-term joint outcomes.

1 | Introduction

After the ankle and knee, the elbow is the most frequently impaired joint in patients with haemophilia [1]. As diarthrodial joint with a significant amount of synovium, elbow is particularly susceptible to this cascade of damaging impact [1, 2]. Understanding clinical and imaging presentations of synovitis and arthropathy of the elbow facilitates accurate diagnosis and applying the most appropriate therapeutic strategies.

Early and continuous haematological prophylaxis (ideally beginning in infancy) is fundamental. Without adequate prophylaxis, persistent joint bleeds can happen in the elbow. Painful haemarthroses will then need aggressive treatment to avert progression to synovitis and eventually haemophilic arthropathy. Without adequate haematological treatment, between the second and fourth decade of life, many patients with haemophilia exhibit painful elbow joint destruction [1–3].

Prior to the advent of effective medical control of haemostasis, on demand treatment or the absence of treatment resulted in compromised target joints including the elbow. Primary and secondary prophylaxis have proven to be effective tools to prevent bleeding, and to slow the progression of arthropathy and its associated impacts [4, 5]. However, the elbow often remains a problematic joint in PWH.

The involvement of an experienced multidisciplinary team allows for a more comprehensive approach and an individualised care, with regular follow-up to optimise compliance and outcomes in people with haemophilia (PWH) [6–9].

The objective of this paper is to present a review of diagnosis and treatment of synovitis and arthropathy of the elbow in haemophilia.

On 1 October 2025, a literature search was conducted in PubMed using the keywords "hemophilia elbow." A total of 466 articles were found, of which 62 were ultimately analysed because they were considered to be directly related to the title of this narrative literature review (inclusion criterion). The rest (404) were not included because they were considered not to meet the aforementioned inclusion criteria. Also three books were reviewed, and information of five chapters was considered of interest for this review.

2 | Discussion

2.1 | Synovitis and Arthropathy: Clinical Presentation and Diagnosis

The clinical presentation of recurrent haemarthrosis of the elbow and synovitis, deserves reflection, given that there are three distinct articulations involved in this joint that contribute to the bleeding pattern, loss of range of motion in different planes and to singular attributes of the arthropathy: (a) the humeroulnar joint, a hinge (ginglymus) joint that permits flexion and extension, (b) the humeroradial joint, a ball and socket type articulation which participates in flexion-extension and acts as a pivot during rotation of the forearm, and (c) the proximal radioulnar joint, a pivot (trochoid) joint between the head of the radius and the radial notch of the ulna, stabilised by the annular ligament [10]. The coexistence of these three distinct functions which produces movement in two different planes and a combination of both while sharing a common joint space, make the elbow a challenging joint for preservation of musculoskeletal health.

The release of haemoglobin and iron from red blood cells that degrade, trigger the molecular pathophysiology of haemophiliac arthropathy [11]. The combination of iron deposition and inflammatory mediators, through different reactions, will lead to synovial hyperplasia and modification on the mechanical chondral properties [12, 13].

The clinical presentation of synovitis of the elbow in haemophilia has an ample spectrum, spanning severe, early, acute limitation of function to imperceptible chronic presence, leading to subclinical joint degeneration. It is the latter that requires attention to create awareness in the modern setting.

Preserving joint integrity has become a lifelong goal in the management of haemophilia. Point-of-care ultrasound is a very useful tool for joint screening, offering up to 40% greater sensitivity than physical examination for detecting synovitis [14, 15]. It enables early identification of haemarthroses, synovial hypertrophy, increased vascularity, and early cartilage changes. Screening tools, such as point-of-care ultrasound, are used to detect disease in the absence of symptoms and thereby increase the likelihood of early intervention. Diagnostic tools, in contrast, identify disease when signs or symptoms are already present, using clinical history, physical examination, and imaging modalities such as ultrasound or MRI. Prognostic tools help predict disease severity and guide personalised management.

To stage the level of involvement of the joint and provide forecasting, magnetic resonance imaging (MRI) is the gold standard. From a diagnostic perspective, synovial hypertrophy will appear iso to hypointense on T1 and hyperintense on T2/PD, constituting the most specific early feature of chronic synovitis. Hemosiderin deposition indicating subclinical or recurrent bleeding will appear as low-signal intensity areas within the synovium on all sequences, most striking on T2 images. Joint effusion will show hyperintense on T2-weighted or PD-fat suppressed images [16]. From the perspective of predicting damage, gadolinium provides avid enhancement of the synovium in the presence of active synovitis, which is a marker of ongoing inflammation. Prior to detection with plain radiographs, MRI can evidence a heterogeneous cartilage signal indicating early structural change. In early synovitis bone marrow oedema and subchondral cysts are absent or minimal. Their appearance marks the transition to haemophilic arthropathy [17].

In the elbow, early signs of arthropathy are equivalent to irreversible loss of range of motion in flexion-extension, pronosupination, or often in both planes. Early detection of subclinical synovitis is the new standard of care. It is of paramount importance to preserve joint function.

2.2 | Rehabilitative Aspects

Rehabilitation programmes have proven effective in treating elbow pathology in PWH. These treatments must be prescribed and adapted on an individual basis depending on the condition of the joint, the haematological treatment, and the patient's age.

In acute injuries, controlling haemostasis, resolving haemarthrosis, and restoring the elbow to its previous condition are essential. However, in chronic injuries, pain relief, functional recovery, and reduction of disability will be the keys to rehabilitation treatment [8, 18–20].

The therapeutic strategies most frequently used in rehabilitation are briefly described below (Table 1).

Individualised therapeutic exercise allows for the restoration of range of motion, progressive strengthening of the elbow flexors/extensors and forearm muscles [21]. It also allows for proprioceptive training and modification of activity, which is essential in cases of instability or ulnar nerve compression [22]. Understanding and controlling pain during exercise is essential for good tolerance [23]. The doses of exercise in the trials varied, but typically spanned several weeks with supervised components and home exercise programmes [18, 20, 21, 24].

Combination of exercises and manual therapy may be used without causing bleeding and pain to increase the functionality,

TABLE 1 | Rehabilitation strategies most frequently used for pathology of the elbow in haemophilia.

Strategy	Characteristics
Therapeutic exercise	Must be individualised, supervised and subsequently maintained at home
Manual therapy	Includes joint traction, fascial therapy, passive muscle stretching and proprioceptive neuromuscular facilitation.
Occupational therapy	Training upper limb function and basic and instrumental activities of daily living
Elbow orthoses	Provide joint protection and progressive recovery of range of motion
Oral analgesia	Paracetamol as first step, escalating to second- and third-level drugs, depending on pain intensity
Intra-articular procedures	Allow local treatment of active synovitis and elbow pain due to haemophilic arthropathy
Educational intervention	Involve patient and family improves motivation and adherence to treatment
Multidisciplinary approach	Coordination with other specialists to ensure a musculoskeletal and biopsychosocial approach to haemophilia

joint health, and quality of life (QoL) [25]. Home exercises may be ameliorated in pain, QoL, and some range of motions (ROM) [26].

Specific manual therapy interventions include joint traction, fascial therapy, passive muscle stretching, and proprioceptive neuromuscular facilitation. These techniques appear to be effective in improving elbow range of motion, arm circumference, and pain in chronic elbow arthropathy in PWH [24].

Occupational therapy focuses on modifying activities, specific training for specific upper limb tasks, and education to improve joint economy in daily activities [27]. It helps patients to maintain, restore or increase their ability to care for themselves, engage in work/school and leisure; develops individual and group programs for all age groups to be as independent and functional as possible in different environments; and enhances physical and psychological wellbeing [28]. It also plays an important role in training the use of technical aids to promote independence in self-care. For patients who need to use crutches, those that are less harmful to the elbow should be recommended. The use of occupational therapy could be effective in improving autonomy and quality of life in haemophilia patients, but more studies are needed [27].

The use of elbow orthoses may be recommended in some specific cases where it may be necessary. The orthoses used for elbows include elastic sleeves, that are adjustable, to provide support and improve proprioception. Static posterior orthoses for immobilising the elbow, which provide mediolateral and rotational stability to the elbow, are usually only prescribed in cases of acute inflammation or severe haemorrhage. Dynamic articulated orthoses for progressively and gradually increasing mobility are also useful in cases of contracture, having been demonstrated to be safe and cost-effective. They may also use to limit flexion or extension to certain degrees in order to prevent the joint reaching vulnerable positions when the aim is to protect against a painful ROM [29] (Figure 1).

Evidence also suggests that orthotic devices improve quality of life and functional outcomes in haemophilia patients with elbow involvement, especially when used to prevent lesions from strenuous activities or to assist in the performance of daily tasks. However, the use of heavy or restrictive orthoses should be minimized to avoid unnecessary immobilization and loss of function [30].

In terms of pharmacological strategies, first-line analgesia usually prioritises paracetamol for pain, with opioids reserved for severe pain in adults when necessary. In the treatment of acute events, a short course of selective cyclooxygenase-2 inhibitors anti-inflammatory agents may also be used. Non-selective non-steroidal anti-inflammatory drugs are not recommended due to the risk of bleeding [7].

Intra-articular injections can also be useful when treating the elbow, as they offer local improvement and avoid the side effects of systemic treatment. The use of radioactive isotopes or chemical agents is a good option in cases of synovitis, as are corticosteroids if there is no other alternative. Platelet-rich plasma (PRP) has also been used as a treatment for chronic synovitis [31]. The use of hyaluronic acid or PRP may be helpful in treating pain from haemophilic arthropathy [32].

Adherence to rehabilitation treatment can sometimes be compromised. Educational interventions aimed at both patients and families are essential to improve understanding and motivation. Therefore, combining rehabilitation therapies and educational sessions seems to be a good way to achieve the best results [24].

It is recommended to assess the response to different therapies through periodic evaluations using validated joint health scores and imaging to monitor progress and adjust rehabilitation protocols [8, 9, 33]. Among the tools the most used ones are Haemophilia Joint Health Score (HJHS) for clinical evaluation, Visual Analog Scale (VAS) for pain, Haemophilia Early Arthropathy Detection with Ultrasound (HEAD-US) for joint damage and Haemophilia Activity List (HAL) and Functional Independence Score in Haemophilia (FISH) for activity and functional impairment [7–9, 33].

Further research is needed to better define best practices in elbow rehabilitation for people with Haemophilia, from a

FIGURE 1 | Different orthoses used in haemophilic elbow: (a) Plain and adjustable elastic orthosis. (b) Immobilising static orthosis. (c) Dynamic jointed orthosis with lockable joints that control or limit joint mobility.

multidimensional care perspective. The involvement of an experienced multidisciplinary team, with the patient and family in the centre of the decision making process allows a more comprehensive approach [6, 8, 9].

2.3 | Synovectomy

In the treatment of the haemophilic elbow, the most commonly used invasive joint preservation technique prior to total elbow arthroplasty is synovectomy. This can be performed alone or in combination with excision of the radial head, in case of severe pain and bleeding in the elbow that occurred in spite of appropriate, episodic, replacement therapy over a period of more than 6 months, associated with hypertrophy of the radial head and a significant loss of pronation-supination [34]. Transposition of the ulnar nerve and removal of heterotopic ossification may be necessary with much less frequency [35, 36].

In the presence of haemophilic synovitis of the elbow causing recurrent haemarthroses, it is reasonable to perform a synovectomy which is achievable by three different methods: by intra-articular injection (synoviorthesis), by arthroscopic surgery, or by open surgery [15].

There are two types of synoviorthesis: radioactive and chemical. Radioactive synoviorthesis (also called radiosynovectomy) is a procedure that has proven to be effective and safe. It can be performed with different radioactive isotopes: chromium phosphate 32P, yttrium-90, rhenium-186, and samarium hydroxyapatite 153. Radioactive synoviorthesis is usually performed with a single injection, with rhenium-186 being the most recommended isotope (Figure 2). After infusion of the isotope, it is recommended to infuse a corticosteroid during needle withdrawal to prevent burns to the periarticular soft tissues [37].

As for chemical synoviorthesis, there are also several publications that have demonstrated its effectiveness. In the literature, some studies indicate that chemical synoviorthesis with rifampicin produces results similar to those of radioactive synoviorthesis in the elbow. The disadvantage of chemical synoviorthesis with rifampicin is that it is highly irritating and painful, and requires several intra-articular injections per week, which may limit the use of this technique especially in younger PWH [38].

In other words, it is unclear in the literature which of the two types of synoviorthesis (chemical or radioactive) is recommended in terms of efficiency. Both approaches are minimally invasive and well-tolerated, but radioactive synovectomy typically provides longer-lasting results and requires fewer procedures, while chemical synovectomy may be preferred in younger children or where radioisotopes are unavailable [37–39]. There are many more publications on radioactive synoviorthesis, which could suggest that most authors prefer radioactive synoviorthesis for the treatment of haemophilic synovitis of the elbow.

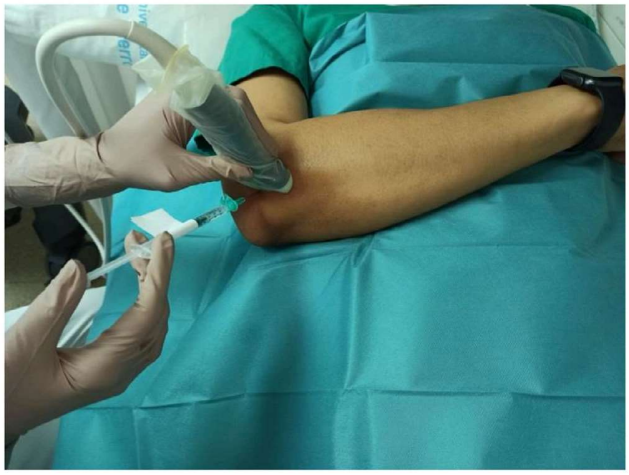

FIGURE 2 | Ecoguided radiosynoviorthesis of the elbow in a patient with haemophilia and active synovtis. Lateral approach to access the radio-humeral joint.

In our experience over a period of 38 years, 500 radioactive synoviorthesis were performed in 443 joints of 345 patients with haemophilia diagnosed with chronic synovitis. The mean patient age was 23.7 years (range, 6–53). The mean follow-up was 18.5 years (range: 6 months–38 years). The RS was carried out with either yttrium-90 or rhenium-186. We performed 1–3 injections, with a 6-month interval between them. RS resulted in significant improvement in all the parameters studied, except in the WFH radiologic score that showed no improvement. On average, the number of haemarthroses decreased by 64.1% and articular pain decreased by 69.4%. The degree of synovitis showed a reduction of 31.3%. The WFH clinical score revealed an improvement of 19%. The WFH radiological score showed no improvement. There were four complications (0.9%) of RS. Twenty-eight (6.3%) joints eventually had to be subjected to arthroscopic synovectomy or total knee replacement. No cancer was observed in this group of

patients during the 38-year period. Radioactive synoviorthesis is an effective, safe, minimally invasive, well tolerated procedure in the long-term for the treatment of chronic haemophilic synovitis. Moreover, it is very easy to perform. The knee required more injections than the elbow or the ankle and more severe synovitis required a higher number of RS procedures [40].

Arthroscopic synovectomy is a surgical procedure in the strict sense, requiring anaesthesia, appropriate surgical instruments, and surgeons specialized in the procedure. Its primary objective is the surgical resection of synovial tissue.

Open surgical synovectomy of the elbow is a less commonly used procedure, although it remains useful when associated with excision of the hypertrophic radial head due to significant loss of prono-supination [34, 41, 42].

When synovectomy of any type fails to control haemarthrosis, the degeneration of the articular cartilage will eventually lead to severe elbow arthropathy. This will cause significant pain and functional disability such that the only solution will be elbow arthroplasty (non-preserving joint surgery). Angiographic embolisation might be considered a therapeutic option when chronic synovitis causes joint haemorrhages that do not respond to replacement of the clotting factor to normal levels [43]. However, is an invasive procedure with a reported rate of complications up to 25% [44].

2.4 | Excision of the Radial Head

Radial head excision in haemophilic elbow arthropathy involves the surgical removal of the radial head, usually accompanied by partial synovectomy. This technique is indicated in PWH who have persistent pain, significant limitation of pronation-supination, and recurrent episodes of joint bleeding, despite adequate replacement therapy and conservative management for at least 6 months, especially when there are hypertrophy and erosion of the radial head that cause mechanical blockage of movement. The literature shows that radial head excision in this context is effective in improving the range of prono-supination (average increases from 63° to 115°), reducing pain (significant improvement on the Visual Analogue Scale), and decreasing the frequency of haemarthrosis [34, 41, 42].

Currently, emphasis is placed on the importance of a detailed preoperative evaluation, especially of the distal radioulnar joint, as its involvement may limit functional benefits after excision of the radial head [41, 42].Technically, it is recommended to preserve the insertion of the annular ligament and avoid excessive resection to minimize the risk of complications [41, 45]. However, the incidence of serious complications, such as significant instability or neuropathy, is low and usually resolve spontaneously.

Recent advances include the use of minimally invasive techniques and the consideration of alternatives such as radial head arthroplasty in selected cases, although in haemophilia, excision of the radial head remains the preferred option due to its lower complexity and risk of prosthetic complications [46, 47].

2.5 | Elbow Arthroplasty

Severe symptomatic haemophilic arthropathy with functional impairment results in joint arthroplasty being the standard of care [4, 48, 49]. On the other hand, not all procedures have the same outcomes in PWH as in the general population. Although knee and hip arthroplasties have demonstrated good clinical results, long-lasting effects, and high levels of patient satisfaction, outcomes of total elbow arthroplasties (TEA) remain a topic of debate [48, 50]. Particularly in the elbow, despite pain reduction and partial (surely not full) recovery of range of motion (flexion), a prosthetic joint required rigid adherence by patients to life-long limitations [50]. The survival rate of elbow implants is not comparable to the hip or knee due to the particular forces acting mostly on the humeral side of the implant during flexion, extension, and weight lifting [48, 50].

When indicated, and once the patient has fully understood these limitations, the prosthesis may be implanted. Several designs during the last decades have been released, as several surgical accesses have been proposed, less invasive than the past [4, 48, 50] (Figures 3 and 4). Cement-less, cemented, or hybrid implants have been adopted with variable outcomes. As usual, the choice depends on the surgeon's ability and familiarity, bone quality (mainly assessed by CT or MRI), age, and functional activity of the PWH [49, 50].

After surgery, the timing of the implementation of rehabilitation as well as the therapeutic techniques considered, is crucial. The need of early range of motion must appreciate associated risks to the surgical site (post-op pain, wound dehiscence, risks of infection and bleeding). Progressively, more involved treatment plans incorporating not only motion recovery, but re-strengthening and proprioceptive training may be implemented [51].

Consistent long-term follow-up data on elbow arthroplasties in PWH is scarce. Regarding prosthetic survival in haemophilia patients, it has been published to be 87.5% at 15 years [52]. In the general population, TEA is a surgical intervention with a high rate of complications (between 16% and 62%) [48]. Complications described include periprosthetic infection, polyethylene wear, and loosening of the humeral component [53] A high rate of need for surgical revision (between 6% and 29%); and with an implant survival at 25 years of 78% [48, 54].

2.6 | Ulnar Nerve Decompression

One peculiar aspect of elbow arthropathy at mid to late stages is ulnar nerve compression in the ulnar tunnel [55]. Osteophyte development at the distal humerus, the proximal ulna, and soft tissue changes impacting flexibility resulting in more rigid characteristics may contribute to the ulnar nerve becoming chronically irritated. Patients usually complain of pain over the ulnar tunnel at the posterior and medial aspect of the elbow, and distal numbness or paresthaesia in the ulnar nerve territory (medial side of the 4^{Th}, 5^{Th} finger). At later stages, motor symptoms arise primarily with adduction of the thumb, abduction and adduction of the digits, and flexion of 4^{Th} and 5^{Th} fingers [36, 4, 49, 55]. Diagnosis is performed by standard x-rays (to assess the presence of osteophytes) and MRI (to evaluate the actual site of

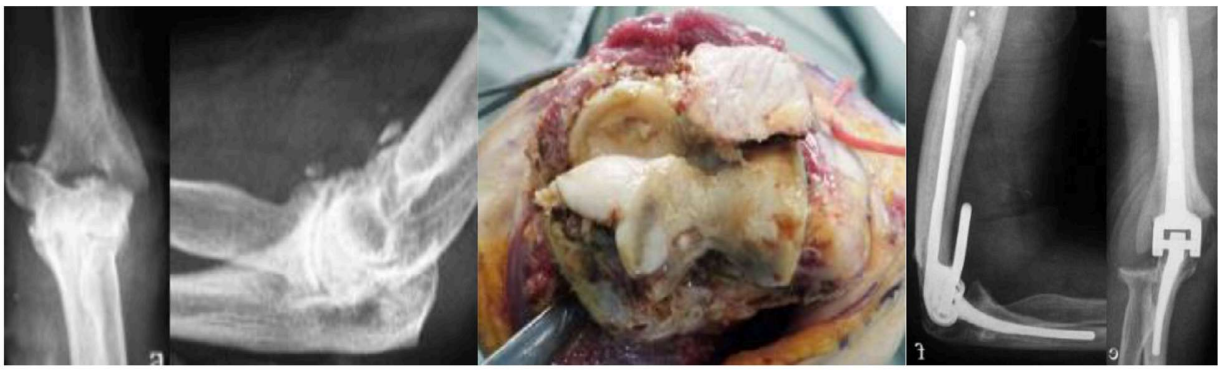

FIGURE 3 | Forty-eight years old male with severe haemophilia A and late stages arthropathy of the right elbow (huge flexion deficit): cemented total elbow arthroplasty.

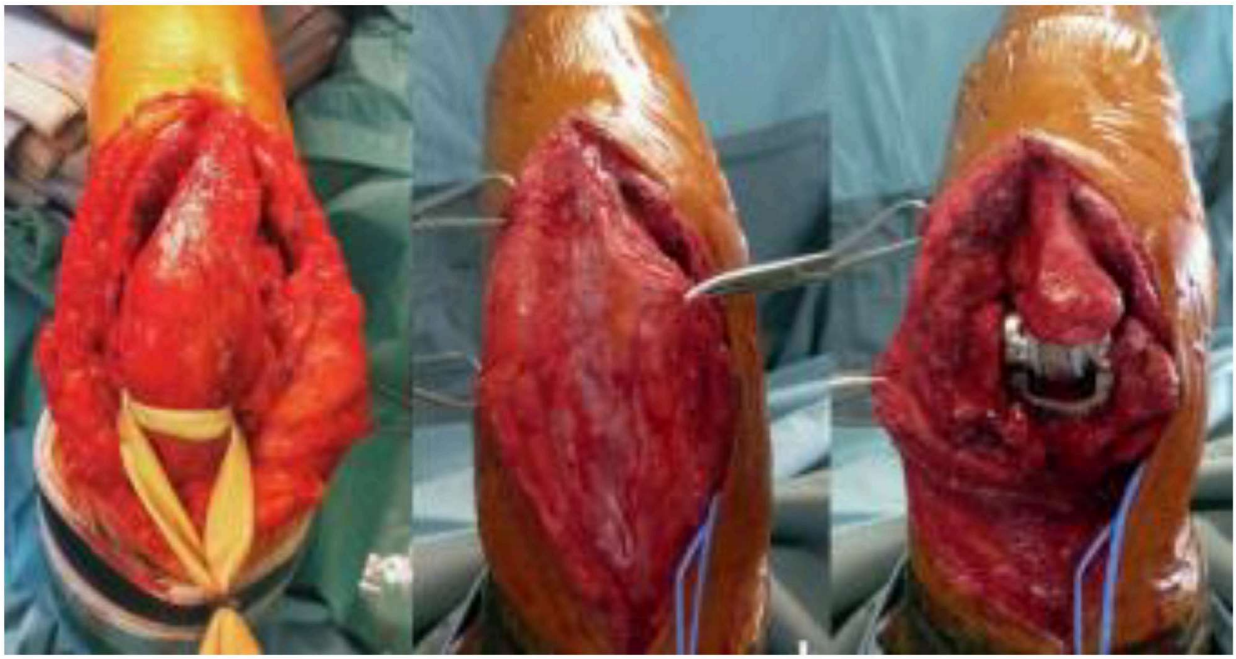

FIGURE 4 | Modern tissue sparing approach for total elbow arthroplasty in a 30-years old male patient with severe haemophilia A and late stages right elbow arthropathy (huge loss of flexion and extension): triceps-on access.

compression of the nerve also in case of soft tissue impingement). Electromyography is useful to verify from a quantitative point of view the entity of the compression, and to perform a differential diagnosis, excluding cervical spine issues. Treatment at the initial stages consists of gentle muscular activity maintenance, neurotrophic drugs, and physical therapy [36, 55]. With worsening of pain and functional impairment, surgery is the only solution. Open ulnar neurolysis and its anteposition is the gold standard of treatment, followed by immobilization in semiflexed casting for 2–4 weeks dependent upon individual surgeons' protocol (Figure 5). After cast removal, gentle physiotherapy is proposed. Outcomes are generally good with expected improvements in the following 2–4 months. This procedure does not directly address range of motion issues, or symptoms arising from the joint itself.

2.7 | Pseudotumors, Fractures, and the Risk of Compartment Syndrome

Pseudotumours and fractures are additional complications that may appear in the haemophilic elbow, and fractures in particular carry a relevant risk of triggering acute forearm compartment syndrome. Haemophilic pseudotumours can present as progressively enlarging masses that may compress nearby neurovascular structures, sometimes producing sensory disturbances or nerve dysfunction. When they cause mechanical or neurological compromise, surgical excision and nerve decompression may be required [56].

Fractures occurring in the context of advanced haemophilic arthropathy can develop spontaneously due to severe osteolysis or

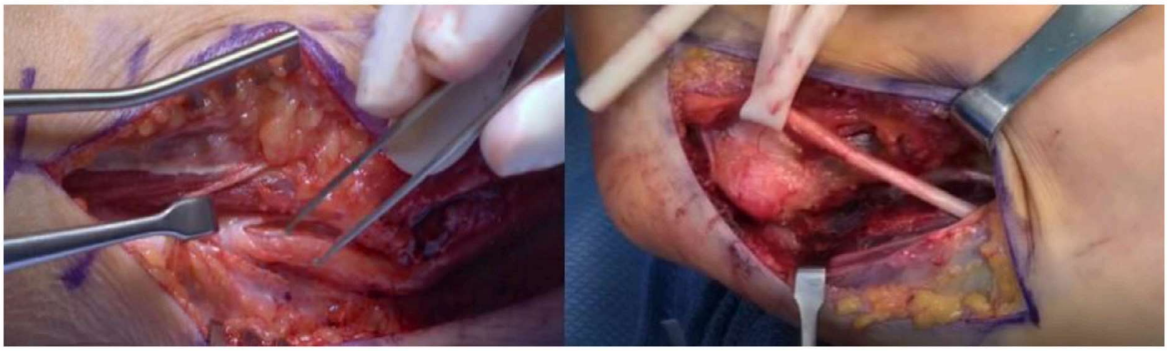

FIGURE 5 | Forty-two years old male with severe haemophilia A undergoing ulnar nerve neurolysis and anterior transposition.

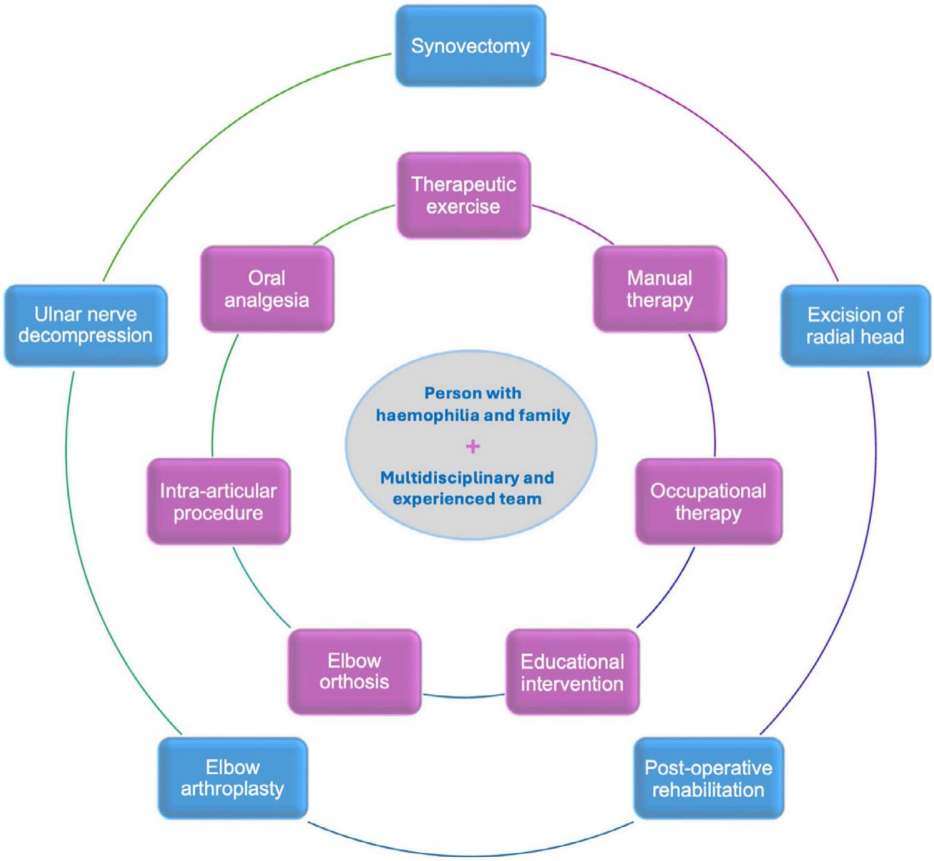

FIGURE 6 | Therapeutic options for synovitis and arthropathy of the elbow in haemophilia: conservative therapies (purple) and surgical procedures (blue).

bone fragility. These injuries may be managed with immobilisation and appropriate factor replacement, and although non-union is possible, patients may still recover acceptable functional levels [57].

In more severe situations, an elbow or forearm fracture may precipitate acute compartment syndrome. Management in this setting demands rapid diagnosis, immediate factor replacement, and emergent fasciotomy to prevent irreversible neuromuscular damage. Delayed recognition—even in individuals with mild haemophilia—can lead to permanent functional loss, neurological deficits, or even limb amputation [58].

2.8 | Post-Operative Rehabilitation in the Elbow

Recovery of function is the primary goal of rehabilitation after elbow surgery in haemophilia, with the caveat that activity levels and appropriate physical demands for the joint may have predetermined limits such as lifting restrictions following TEA [50, 54].

Pre-operative therapy should be considered to optimize soft tissue function prior to the procedure which may improve final outcomes [59, 60]. It may also acclimatize the patient to implementation of the therapy program after surgery and

provide opportunity to have questions answered and information clarified. It supports a crucial element of having an established baseline prior to surgery against which post-operative evaluation may be compared.

Post-operatively it is imperative that pain and swelling be addressed in the inflammatory stage of healing as the former inhibits motivation to mobilize, and the latter if not managed is a precursor to increased scar formation [60, 61], which may have lasting deleterious effects on recovery. Techniques such as limb elevation, compression garmenting, gentle active ROM and positional splinting may be useful and should be operationalized in concert with other strategies such as medical pain management [62].

Specific protocols backed by solid literature are lacking. Rehabilitation after synoviorthesis of the elbow in haemophilia is typically less intensive and allows for earlier return to activity (24–48 h), followed by prompt initiation of gentle range-of-motion and strengthening exercises. Rehabilitation after surgical synovectomy requires a longer period of rest, more cautious and gradual progression of mobilization to prevent stiffness and restore function and a greater emphasis on muscle strength from weeks 3 to 6 [63].

There are gaps in the specific rehabilitation guidelines for other procedures. Elbow radial head excision requires early progressive pronation-supination mobilisation during the first week and strengthening from week 4. Arthroplasty in haemophilia requires immediate active-assisted movement, avoiding end-range stress and very gradual strengthening, with lifelong weight-bearing restriction (≤2–5 kg). Except after synoviorthesis, rehabilitation programmes may last 8–12 weeks and must be carried out ensuring adequate factor replacement to minimize perioperative bleeding risk [9].

Research has shown that low load prolonged stretch (LLPS) and total end range time (TERT) are most effective to address ROM loss, making static and dynamic splint designs an effective complement to active mobility as early on as possible [27, 64]. Techniques once popular such as continuous passive motion are not supported by convincing evidence as a means to offer long term solutions [65, 66], and should be replaced by exercise focusing on active control as well as LLPS and TERT.

One component of baseline pre-operative assessment that may be difficult to quantify dependent upon joint health status, is proprioception. Impaired joint position sense, neuromuscular control and movement coordination of the elbow, make targeted proprioceptive retraining an essential component of rehabilitation to optimize joint stability and functional outcomes after elbow surgery in patients with haemophilia [22].

More commonly considered when addressing lower extremity rehabilitation due to hip and ankle strategies of balance control, kinaesthetic sense recovery around the elbow can be critical to optimising function of the upper limb [67].

The elbow is an important partner to the shoulder, and less so the wrist, in performing placement of the hand for task completion. With tasks requiring the hand to be outside of the visual field, mechanoreceptor mediated control of position allows increased efficiency of movement, and overall energy conservation important to long term post-operative capacity.

Historically, and still in many cases, elbow surgery in haemophilia is a salvage procedure to manage end-stage arthropathy and pain [50, 54]. As non-factor therapies, re-balancing agents, and new generation factor products enter more wide-spread usage in resource-available jurisdictions, acute injuries such as distal bicep and collateral ligament tears, fractures of the distal humerus and radial head, and repetitive strain conditions around the elbow may begin to present more frequently in the bleeding disorders population. It is therefore recommended that therapists on comprehensive care teams be familiar with associated protocols, and ready to plan rehabilitation for patients operating at much higher baseline levels of function than in years past, when surgery is not performed for primary consequences of prior joint bleeding.

A footnote to any discussion of therapy post-surgery must focus on formal tracking of outcomes, and a call for consistency in approach and tools employed by practitioners to evaluate change markers. Small cohorts of haemophilia patients, reduced further by subsets undergoing elbow surgery, hinder the process of developing research data sufficiently powered to draw global conclusions. Whether disease-specific measures are utilised, or validated general functional measures of elbow joint health are chosen, sharing data and ensuring comparability of data sets from one jurisdiction to another is imperative.

3 | Conclusion

The elbow is a well-known target joint in people with haemophilia and deserves specific attention and treatment. Understanding clinical and imaging presentations of synovitis and arthropathy is paramount to selecting the best therapeutic options. It is reasonable to adopt the most conservative or less invasive procedure in order to reduce patients' symptoms and restore joint function. Current knowledge supports the combination of multimodal approaches (Figure 6). Prophylaxis reduces factors that trigger bleeding and rehabilitation improves range of motion, strength, proprioception and levels of pain. Synovectomy treats persistent synovial disease, and surgical techniques such as radial head resection, arthroplasty or ulnar nerve release seek to improve function and reduce pain supported by a post-operative rehabilitation programme. The ultimate goal is to improve upper limb function, and quality of life for people with haemophilia.

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Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

The authors have nothing to report.

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