Eosinophilic Fasciitis in Pediatric Patients: A Rare but Distinct Autoimmune Fibrosing Disorder

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PRACTICE GAPS/EDUCATION GAPS

Eosinophilic fasciitis (EF) is a rare, underrecognized fibrosing disorder in pediatric patients, often misdiagnosed or diagnosed late because of its heterogeneous presentation and lack of standardized diagnostic criteria. There is a need to increase awareness of early diagnostic strategies, treatment algorithms, and long-term management considerations to prevent irreversible morbidity in children with EF.

OBJECTIVES *After completing this article, readers should be able to:*

1. Name the clinical features of EF in pediatric patients and how they differ from adult cases.
2. Differentiate EF from other autoimmune or fibrosing disorders using clinical, laboratory, imaging, and histologic findings.
3. Describe current evidence-based treatment strategies for pediatric EF, including corticosteroids, immunosuppressants, and rehabilitative interventions.
4. Describe the long-term sequelae and prognosis of EF in children and identify areas requiring further research.

ABSTRACT

Eosinophilic fasciitis (EF), or Shulman syndrome, first described in 1974, is a rare fibrosing disorder characterized by painful, symmetric swelling and progressive woody induration of skin and subcutaneous tissues. Although the pathogenesis remains unclear, EF is considered immune-mediated, often triggered by physical exertion, infections, or medications. Pediatric EF, a particularly uncommon subset, can exhibit distinct clinical features, including pronounced extracutaneous manifestations, unpredictable disease progression, and variable therapeutic responses. Clinical presentations range from rapidly advancing fibrosis leading to joint contractures to fluctuating inflammatory episodes. Diagnosis is challenging because of the absence of universal criteria, although peripheral eosinophilia, elevated

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ABBREVIATIONS

EF eosinophilic fasciitis
MRI magnetic resonance imaging

inflammatory markers, and imaging findings support clinical suspicion. Definitive diagnosis depends on deep skin and fascial biopsy, revealing eosinophil-rich lymphoplasmacytic infiltrates and fibrosis. Management primarily relies on systemic corticosteroids, supplemented by steroid-sparing immunosuppressive medications in refractory cases. Early diagnosis and treatment are critical because untreated EF can cause irreversible fibrosis and significant functional impairment. The disease's rarity, heterogeneous presentations, and unclear etiology further complicate clinical management. Recent insights suggest EF may involve intricate interactions among environmental triggers, immune dysregulation, and fibrotic remodeling. This review aims to provide an updated overview of pediatric EF, highlighting current knowledge on clinical manifestations, diagnosis, differential diagnosis, therapeutic approaches, and outcomes, supported by an illustrative case, with emphasis on areas needing further research.

INTRODUCTION

Eosinophilic fasciitis (EF), also referred to as Shulman syndrome, was first described in 1974 by Dr Lawrence Shulman, a Johns Hopkins rheumatologist who later became the inaugural director of the National Institute of Health's Institute of Arthritis and Musculoskeletal and Skin Diseases.[1,2] This rare connective tissue disorder is characterized by painful, symmetrical swelling and progressive woody induration of the skin and subcutaneous tissues.[3] Although originally described as a fibrosing disorder of the fascia, accumulating evidence suggests EF is an immune-mediated condition with a complex interplay of environmental triggers, immune dysregulation, and fibrotic remodeling.[4] The etiology remains unclear, but proposed triggers include physical exertion, infections, and certain medications.[4,5] Since its original description, approximately 300 cases of EF have been reported in the literature, with most occurring in individuals of white or European descent.[6] EF primarily affects adults in their 40s and 50s, although pediatric cases have been reported less frequently.[6,7] Pediatric EF, a small subset of the broader EF population, can display unique clinical features, such as more pronounced extracutaneous symptoms and a variable response to therapy.[6,8,9]

The disease course in children can be unpredictable, with some cases demonstrating a rapidly progressive fibrotic phase leading to joint contractures, whereas others exhibit fluctuating inflammatory symptoms.[10] Laboratory findings, including peripheral eosinophilia and elevated inflammatory markers, may aid in diagnosis but are not universally present.[11] Deep skin and fascial biopsy remain the gold standard for confirmation, revealing lymphoplasmacytic infiltrates with eosinophils and fibrosis.[12] Although spontaneous remission can occur, treatment relies on systemic corticosteroids, often supplemented with immunosuppressive agents in refractory cases.[6] This article provides an overview of the current understanding of pediatric EF, including diagnostic criteria, clinical presentation, differential diagnosis, therapeutic strategies, and an illustrative case.

BACKGROUND

Pathophysiology

The precise mechanisms of EF remain unknown. EF is believed to result from immune dysregulation leading to eosinophilic and lymphocytic infiltration of the fascia.[4,6] The presence of peripheral eosinophilia suggests a role for eosinophil-mediated inflammation, with activated eosinophils releasing toxic granules that contribute to tissue damage and fibrosis.[13] Inflammatory cytokines, including interleukin (IL)-4 and IL-5, promote eosinophil recruitment and persistence in affected tissues.[13–15] transforming growth factor-β plays a central role in fibroblast activation, leading to excessive collagen deposition and progressive fascial thickening.[16]

Histopathological examination of EF-affected tissues typically reveals fascial thickening with inflammatory infiltrates composed predominantly of eosinophils and lymphocytes.[17] The early stages typically reveal edema in the deep fascia and lower subcutaneous tissue, accompanied by perivascular and interstitial infiltration of lymphocytes, plasma cells, histiocytes, and eosinophils.[18] As the disease advances, there is increased fibroblast proliferation and dense collagen deposition. Fibroblast activation is a hallmark of EF pathogenesis, contributing to excessive extracellular matrix deposition and eventual fibrosis of the fascia and surrounding structures.[13] Unlike systemic sclerosis, EF does not exhibit significant vascular involvement or autoantibody production. This may reflect EF's distinct pathophysiology, which lacks the widespread endothelial injury and autoantibody-mediated mechanisms seen in systemic autoimmune diseases.[19,20] Magnetic resonance imaging (MRI) reveals fascial thickening and hyperintensity on T2-weighted sequences" So that the preceding phrase defines T2-weighted sequences as a type of readout for MRI.[21] Over time, chronic inflammation leads to irreversible fibrosis, causing skin retraction, joint contractures, and long-term functional impairment (Figure 1). In one of the largest early EF case series, over 50% of patients developed joint contractures, although the proportion requiring orthopedic intervention was not reported.[22]

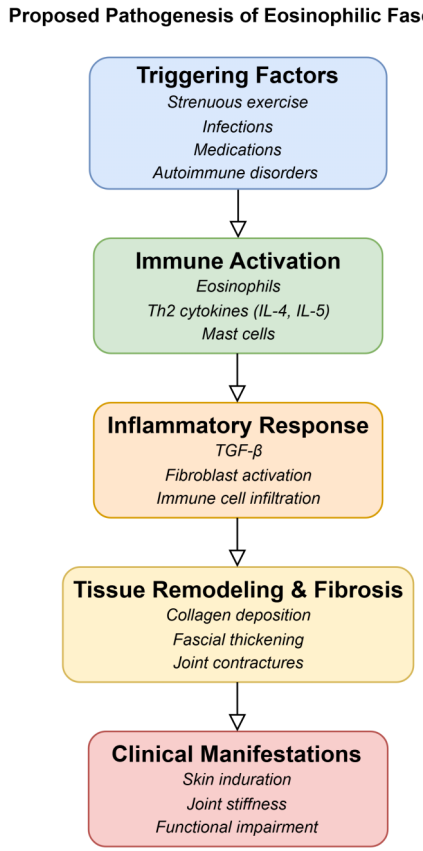

FIGURE 1. Proposed pathophysiology of EF in pediatric patients. Abbreviation: EF, eosinophilic fasciitis.

Triggers

Several environmental and systemic factors have been implicated in the development of EF. Strenuous physical activity, such as intensive athletic training or repetitive exertion,[23,24] often precedes symptom onset, particularly in adolescent patients, suggesting that repetitive mechanical stress may trigger an abnormal immune response in predisposed individuals.[5,6] Certain medications, including statins (eg, simvastatin and atorvastatin), immune checkpoint inhibitors (eg, nivolumab, pembrolizumab, and atezolizumab) proton pump inhibitors (eg, lansoprazole and esomeprazole), and antiepileptic drugs, have been associated with EF onset, although the mechanism is unknown.[4,17] Rare cases have been linked to supplements like L-tryptophan,[25] and blood disorders such as aplastic anemia and multiple myeloma.[26,27] Some cases of EF occur in the context of autoimmune diseases such as juvenile idiopathic arthritis, Sjögren syndrome, or systemic lupus erythematosus, suggesting that underlying immune dysfunction may predispose individuals to the development of this disorder.[7,28,29]

Infections, particularly viral or bacterial illnesses, have been identified as common antecedents.[6,8] *Borrelia burgdorferi* has been associated with EF in multiple cases.[30,31] Additionally, a case of SARS-CoV-2 has been reported as a trigger.[32] Another documented case suggests a possible link between EF and parasitic infections, in which a patient with intestinal parasitosis due to *Ascaris lumbricoides* developed EF-like symptoms, including palpebral edema and marked eosinophilia. Despite antiparasitic treatment, symptoms persisted, and a diagnosis of EF was confirmed via skin and muscle biopsy.[33] The mechanism here could involve molecular mimicry or immune system priming, leading to abnormal inflammatory responses targeting the fascia. However, given that most reports are case studies, the association remains anecdotal because no definitive causative pathogen has been identified. Additionally, whereas infections such as borrelia or mycoplasma have been reported in isolated cases, these triggers appear less common than medication exposures or physical exertion, which are more frequently implicated in EF pathogenesis.[5,6]

Morphology

Pediatric EF primarily manifests as symmetrical, indurated plaques overlying the distal extremities.[6,31] In the early inflammatory phase, affected areas may appear edematous or erythematous, mimicking cellulitis or deep venous thrombosis.[4] As the disease progresses, the skin adopts a characteristic "peau d'orange" texture due to underlying fascial fibrosis, with firm, immobile plaques and exaggerated skin folds that may spread proximally. The progressive thickening of the fascia leads to restricted joint mobility and contractures, particularly in the wrists, ankles, and knees.[5,6] This predilection may relate to the high mobility and frequent mechanical use of these joints.

Although the classic presentation of EF is relatively well recognized, atypical forms of the disease have been described. Some cases present with solitary plaques or nodules rather than the classic fibrotic thickening.[34,35] Unlike systemic sclerosis, EF does not typically trigger Raynaud phenomenon, which does occur occasionally in children, or significant internal organ involvement, which aids in distinguishing it from other connective tissue disorders.[4,6] Epidermal and dermal atrophy are typically absent.[18] In pediatric cases, extracutaneous manifestations such as myositis, arthritis, and hematologic abnormalities, including peripheral eosinophilia, hypergammaglobulinemia, and elevated inflammatory markers, are largely similar to those in adults but may be more pronounced, adding complexity to diagnosis and management.[7,22,36] However, some case reports have described mild interstitial lung disease and esophageal dysmotility in patients with EF, raising the possibility of subclinical visceral involvement in a small subset of cases.[37]

DIAGNOSTIC CRITERIA

The diagnosis of EF is primarily clinical but requires a combination of characteristic physical findings, laboratory abnormalities, imaging studies, and histopathological confirmation. Common physical signs include painful symmetric swelling, skin induration with a woody texture, a peau d'orange appearance of the skin, and the groove sign, which refers to linear depressions following superficial veins caused by fascial tethering.[6] Unlike systemic sclerosis, EF lacks well-established classification criteria, making early recognition challenging, particularly in pediatric patients. Deep skin and fascial biopsy is the gold standard for definitive diagnosis, typically showing fascial thickening and fibrosis, with or without a lymphocytic infiltrate. Although tissue eosinophilia may be present, it is not required for diagnosis. Additional diagnostic modalities aid in the clinical suspicion and assessment of disease severity.[38]

Peripheral eosinophilia is present in over 60% of cases, although it is not a mandatory diagnostic feature.[5,6] Other laboratory findings may include hypergammaglobulinemia, elevated erythrocyte sedimentation rate, and increased C-reactive protein, reflecting systemic inflammation.[6] Autoantibody testing, including antinuclear antibodies, is typically negative,[19,20] helping to distinguish EF from systemic autoimmune diseases such as juvenile systemic sclerosis or dermatomyositis. Additionally, increased expression of soluble CD40 ligand (sCD40L) has been reported in EF, with significantly higher serum levels compared with healthy controls.[39] sCD40L levels normalized following corticosteroid treatment, suggesting a potential role as a disease activity marker, although further studies are needed to confirm sCD40L's clinical utility.

MRI is a noninvasive tool in EF diagnosis, especially for assessing the extent of fascial involvement. Typical MRI findings include fascial thickening, T2 hyperintensity indicating edema and inflammation, and enhancement with gadolinium contrast.[6] These findings correlate with active disease and can be used to monitor treatment response. Ultrasonography has also been explored as an adjunct modality,[40] demonstrating increased echogenicity and thickening of the fascia, and decreased subcutaneous compressibility.[38,41] Compared with MRI, ultrasonography is less invasive, does not carry the potential need for sedation in pediatric patients, and can be performed at the bedside or in outpatient settings, making it particularly useful for initial screening and treatment monitoring in children.

Clinically, EF is characterized by symmetrical, indurated skin thickening, predominantly affecting the extremities. Early in the disease course, the skin may appear edematous, but as fibrosis progresses, it develops a "woody" consistency with a peau d'orange texture. Joint contractures are a late-stage complication due to fascial tightening. Unlike systemic sclerosis, EF does not involve digital ulcers, or internal organ fibrosis, which are key distinguishing features.[4]

Because of the absence of universally accepted diagnostic criteria, a combination of clinical assessment, laboratory findings, imaging, and histopathology is used for confirming EF (Figure 2). Early recognition can prevent irreversible fibrosis and functional impairment, particularly in pediatric patients, for whom the disease course can be unpredictable. Although onset may be abrupt, developing within days to weeks,[8] progression to fibrosis and joint contractures can occur over weeks to months,[42] stressing the importance of timely diagnosis and intervention.

ASSOCIATIONS

As mentioned, EF has been linked to a range of systemic conditions, environmental triggers, and underlying immune dysregulation, although exact etiology is unknown.[13,43] Several cases have identified associations between EF and autoimmune diseases, hematologic disorders, and malignancies, as well as medication exposures such as immune checkpoint inhibitors, which are cancer immunotherapies that enhance T-cell activation by blocking inhibitory pathways; reported examples include nivolumab and pembrolizumab.[27,44–47] These associations suggest that EF may be a

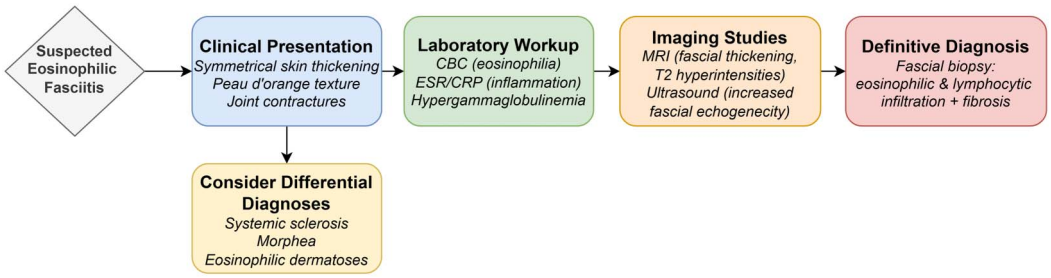

FIGURE 2. Diagnostic approach to EF.
Abbreviation: EF, eosinophilic fasciitis.

heterogeneous disorder with multiple contributing factors rather than a singular pathogenic mechanism.

One of the most commonly reported associations is with autoimmune diseases. Studies have documented cases of EF occurring alongside conditions such as myeloproliferative disorders, systemic lupus erythematosus, Sjögren syndrome, and autoimmune thyroiditis.[8] The presence of overlapping autoimmune features suggests that EF may be part of a broader immune dysregulation spectrum, although autoantibody production is absent. Although EF is not classified as a systemic autoimmune disease, its response to immunosuppressive therapy supports the hypothesis of immune-mediated pathogenesis.

Rarely, EF has been linked to hematologic malignancies, with a scoping review finding EF being associated with paraneoplastic syndromes in 10% of cases.[27,48] Although rare in pediatric cases, EF has been associated with hematologic malignancies such as chronic lymphocytic leukemia, non-Hodgkin lymphoma, and multiple myeloma in adult populations.[17,27,49] The mechanism linking EF to these conditions remains speculative, but paraneoplastic processes involving cytokine dysregulation and aberrant immune activation have been proposed.[50]

TREATMENT

The management of EF is largely empirical because no standardized treatment guidelines exist because of the rarity of the disease, especially in pediatric populations. Given the complexity of EF and the need for immunosuppressive therapy and coordinated rehabilitation, pediatric patients should be managed by a multidisciplinary team, typically led by a pediatric rheumatologist in collaboration with dermatology and physical therapy. The primary goal of therapy is to reduce inflammation, prevent fibrosis, and preserve joint mobility. Treatment approaches are typically divided into first-line therapies, which focus on rapid disease control, and second-line therapies, which are reserved for refractory cases or for reducing long-term corticosteroid dependence.

First-Line Therapies

Systemic corticosteroids are the cornerstone of EF treatment and are the first-line therapy in both pediatric and adult patients. High-dose prednisone (1–2 mg/kg/d) is the most commonly used initial regimen, with gradual tapering over several months to prevent disease relapse.[5,6] Corticosteroids effectively reduce inflammation and improve skin induration in the early inflammatory phase of the disease in up to 70% of adult cases.[51] In pediatric EF, recent case series also support the effectiveness of corticosteroids as part of combination regimens, with most children achieving clinical improvement or remission when treated early and alongside methotrexate.[7] However, their efficacy in reversing established fibrosis is limited, which stresses the importance of early intervention.

In addition, physical therapy is crucial in EF management, particularly in preventing joint contractures and preserving range of motion. Because of fascial thickening and fibrosis, children often develop stiffness in the wrists, elbows, and ankles, which can significantly impact mobility. A structured rehabilitation program, including stretching exercises and occupational therapy, is recommended early in the disease course to minimize functional impairment. Case-based evidence supports the use of multidisciplinary regimens that include stretching, strengthening, massage, splinting, and occupational therapy, with 1 report documenting marked improvement in range of motion and function without exacerbation of disease activity.[52] Despite this, one of the largest patient cohorts analyzing EF management found only 37% of patients being referred for physical therapy.[53]

Second-Line Therapies

For patients who do not respond adequately to corticosteroids or who experience significant side effects, steroid-sparing immunosuppressive agents are employed as second-line therapies. Methotrexate is the most frequently used immunosuppressant and has shown efficacy in reducing steroid dependence while maintaining disease control.[4,54] Mycophenolate mofetil has also been used in refractory cases or in combination with corticosteroid to maintain disease suppression, particularly in patients with extracutaneous involvement, although data on its use in pediatric EF is limited.[55–57]

Other immunosuppressive agents, such as azathioprine, cyclosporine, cyclophosphamide, and hydroxychloroquine, have been reported in case series with variable success.[22,31,58] However, spontaneous recovery in some patients makes it challenging to assess its efficacy definitively. Further, most of the known cases are from the adult population. Recently, biologic therapies have been explored, including rituximab and intravenous immunoglobulin, particularly in steroid-refractory EF.[59,60] These agents are experimental, and their long-term efficacy in pediatric EF has yet to be fully established.

Adjunctive therapies such as nonsteroidal anti-inflammatory drugs and colchicine can be used to manage pain and inflammation in milder cases. These drugs are insufficient as monotherapy for active disease.[61,62] Antifibrotic agents, including D-penicillamine and pirfenidone, have been proposed for preventing irreversible fibrosis.[62–64] In an adult

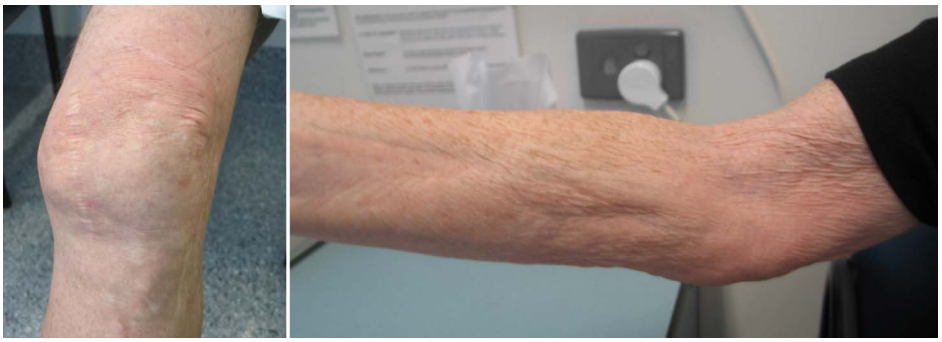

FIGURE 3. Cutaneous and musculoskeletal sequelae of pediatric EF.
Abbreviation: EF, eosinophilic fasciitis.

study comparing treatment regimens, patients receiving D-penicillamine plus corticosteroids showed significantly greater clinical improvement compared with those receiving corticosteroids alone[65]; however, although D-penicillamine was effective, it was associated with adverse effects such as proteinuria.

Ultimately, treatment strategies for pediatric EF should be individualized based on disease severity, response to therapy, and the presence of extracutaneous manifestations. Early initiation of corticosteroids remains the most effective approach for preventing fibrosis, whereas second-line immunosuppressive agents are used for maintaining long-term disease control in refractory cases.

CLINICAL SNAPSHOT: LONG-TERM SEQUELAE OF PEDIATRIC EF

A patient aged 21 years presented with a history of EF diagnosed at aged 16 years, with no identifiable triggers at the time of onset. The patient was left with persistent cutaneous and musculoskeletal sequelae, including significant atrophy of the soft tissue, skin induration in the distal upper extremity, and mild flexion contractures of the elbow (Figure 3). Functional limitations included reduced range of motion, difficulty with full elbow extension, and discomfort with repetitive upper limb movements.

DISCUSSION AND FUTURE DIRECTIONS

Because of the rarity of pediatric EF and its overlap with other connective tissue disorders, diagnosis may be delayed and the condition underrecognized, particularly as juvenile patients less frequently exhibit classic cutaneous signs seen in adults, such as the groove sign or peau d'orange.[7] Despite sharing some clinical and histopathologic features with systemic sclerosis and other autoimmune diseases, EF is a distinct entity with unique pathophysiological mechanisms and a variable disease course. The lack of standardized diagnostic criteria and treatment guidelines presents challenges in clinical practice, contributing to delays in diagnosis and heterogeneity in management approaches.

One of the key challenges in EF diagnosis is the absence of a definitive biomarker. Although peripheral eosinophilia, hypergammaglobulinemia, and elevated inflammatory markers are frequently observed, they are not universally present and may fluctuate throughout the disease course. Histopathologic evaluation remains the gold standard for diagnosis.[12] However, deep fascial biopsy is invasive and may not always be conclusive. Imaging techniques such as MRI have emerged as useful noninvasive tools for assessing fascial involvement.[21] However, their role in routine diagnosis and disease monitoring needs further validation. The development of more sensitive and specific biomarkers for EF would significantly enhance diagnostic accuracy and allow for earlier therapeutic intervention.

From a treatment standpoint, systemic corticosteroids are the first-line therapy for EF, with immunosuppressive agents such as methotrexate or mycophenolate mofetil serving as steroid-sparing alternatives.[54,56] However, treatment response varies among patients, and refractory cases present a major clinical challenge. Although biologic therapies such as rituximab and intravenous immunoglobulin have shown promise in steroid-refractory EF,[59,60] studies into their long-term efficacy and safety are largely preliminary, especially in pediatric populations. There is a growing need for clinical trials evaluating targeted immunomodulatory therapies, particularly in pediatric patients for whom treatment data are even more limited. Exploring antifibrotic agents that could mitigate irreversible tissue remodeling in EF is another promising area for future research.

The etiology of EF remains elusive, although immune dysregulation, environmental triggers, and mechanical

stress have been implicated.[4,5] Future studies should aim to delineate the molecular mechanisms driving inflammation and fibrosis in EF, particularly the role of cytokines, fibroblast activation, and extracellular matrix remodeling. Given the observed associations between EF and other autoimmune diseases, investigating potential genetic predispositions and immune system dysregulation would be useful in further uncovering pathogenesis.

Longitudinal studies assessing long-term outcomes in pediatric patients with EF are also needed. Although some children experience spontaneous remission, others progress to chronic fibrotic disease with significant functional impairment. Identifying early predictors of disease severity and treatment response would help tailor therapeutic strategies to individual patients. Additionally, incorporating patient-reported outcomes and quality of life assessments into research studies will provide a comprehensive understanding of the true burden of EF in pediatric populations. International registries could facilitate the collection of larger patient cohorts.

In conclusion, although significant progress has been made in recognizing and managing pediatric EF, many unanswered questions remain regarding its pathogenesis, optimal treatment strategies, long-term outcomes, and true disease burden. Future research should prioritize the development of standardized diagnostic criteria, identification of novel therapeutic targets, and longitudinal studies to better define disease prognosis.

Summary

- Early recognition and biopsy-confirmed diagnosis of pediatric EF is essential to prevent irreversible fibrosis and functional impairment. (Level C/D: Based on consistent findings from multiple observational studies and expert consensus, delayed diagnosis can result in joint contractures and lasting morbidity.[5,6,12])
- MRI is a valuable, noninvasive diagnostic tool that identifies fascial thickening and active inflammation, often guiding the decision for biopsy and tracking response to therapy. (Level B: Imaging features have shown consistency across studies and are recommended for disease assessment.[21,38])
- Systemic corticosteroids remain the first-line treatment and are effective in reducing inflammation and halting disease progression in most patients. (Level B: Multiple case series and retrospective cohort studies support their early use.[5,6,51])
- Steroid-sparing agents such as methotrexate and mycophenolate mofetil should be considered in refractory or relapsing cases to minimize long-term steroid exposure. (Level B: Based on observational data and practice guidelines in rheumatology.[54,56,57])
- Physical and occupational therapy are underutilized despite their critical role in preserving range of motion and preventing contractures. (Level C: Observational evidence suggests only 37% of patients are referred, indicating a gap in practice.[53])
- Biologic agents (eg, rituximab, IVIG) and antifibrotics (eg, D-penicillamine) are promising for steroid-refractory EF but lack high-quality evidence in pediatric populations. (Level D: Current recommendations are based primarily on case reports and expert opinion.[59,60,65])
- There is a pressing need for standardized diagnostic criteria and prospective studies in pediatric EF to guide treatment and improve outcomes. (Level D: This statement is based on the absence of randomized control trials and guideline consensus.)

Take the quiz! Scan this QR code to take the quiz, access the references and view and save images and tables (available March 1, 2026).

PIR QUIZ

1. A pathologist receives a deep skin and fascial biopsy tissue sample from a patient with peripheral eosinophilia and elevated inflammatory markers. The pathologist reports that the histopathologic findings on this sample are suggestive of EF. Which of the following is considered the hallmark pathophysiologic finding in EF?

   A. Antibody-mediated complement deposition.
   B. Endothelial cell swelling and vasculitis.
   C. Fibroblast activation leading to extracellular matrix deposition and fibrosis.
   D. Macrophage activation and granuloma formation.

2. In the patient in the above vignette, which of the following diagnostic modalities is considered the gold standard for confirming the diagnosis of EF in this patient?

   A. Blood testing.
   B. Computed tomography.
   C. Histopathology.
   D. Ultrasonography.

3. A 9-year-old boy who is being treated for EF is brought to the clinic for possible relapse. The patient was diagnosed with EF at 8-and-a-half years old and has been well-controlled on therapy. In monitoring response to treatment, which of the following findings support the diagnosis of active EF disease and best help assess the extent of fascial involvement?

   A. Erythematous, peeling rash over large joints.
   B. MRI showing fascial thickening with gadolinium enhancement.
   C. Positive antinuclear antibody testing.
   D. Superficial skin biopsy showing macrophages.

4. A 12-year-old girl has been recently diagnosed with EF. The clinician discusses with the parents the treatment options including treatment duration, side effects, and efficacy. Which of the following treatment regimens is considered first-line therapy for EF in this patient?

   A. Botulinum toxin injections.
   B. Intravenous immunoglobulin.
   C. Systemic corticosteroids.
   D. Topical tacrolimus.

5. The patient in the above vignette is started on systemic corticosteroids and methotrexate combination therapy. The clinician discusses with the parents the treatment regimen and its side effects. In addition to the above treatment regimen, which of the following is considered a key complement to steroid therapy to preserve mobility in this patient?

   A. Azathioprine.
   B. Colchicine.
   C. Hyperbaric oxygen.
   D. Physical therapy.

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