Parsonage-Turner Syndrome: An update

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Abstract

Parsonage-Turner Syndrome (PTS), known as Neuralgic Amyotrophy or Brachial Plexus Neuritis, is a rare neurological disorder characterized by sudden onset shoulder pain followed by muscle weakness and atrophy. Despite advancements in medical research, the absence of standardized diagnostic and treatment protocols poses significant challenges. This review explores the multifaceted aspects of PTS, including its genetic predispositions, diagnostic advancements, treatment efficacy, and the long-term impacts on patients' quality of life. The heterogeneity of PTS presentations, the lack of specific biomarkers, and the unclear etiology contribute to the complexities in managing this condition. While current treatments vary from conservative to aggressive interventions, their efficacy remains inconsistent due to the syndrome's unpredictable prognosis. The disease's social and psychological impacts necessitate a comprehensive, multidisciplinary approach to patient care. This review underscores the need for further research to elucidate PTS's genetic and immunological underpinnings, develop more accurate diagnostic tools, and establish evidence-based treatment guidelines.

Keywords: Brachial Plexus Neuritis; Parsonage-Turner Syndrome; Neuralgic Amyotrophy; Physiopathology; Diagnosis; Therapy.

1. Introduction

Parsonage-Turner Syndrome (PTS), or neuralgic amyotrophy or brachial plexus neuritis, is a rare peripheral nerve disorder characterized by sudden, severe shoulder and upper arm pain followed by marked muscle weakness or atrophy1.

This syndrome has been a subject of medical scrutiny due to its abrupt onset and the significant impact it can have on the quality of life. Despite its identification in the mid-20th century, the pathogenesis, optimal diagnostic strategies, and treatment protocols for PTS remain under continuous exploration and debate2,3.

The incidence of PTS is reported to be about 1-3 cases per 100,000 people annually, affecting men more frequently than women and showing no clear preference for any specific age group4.

However, a bimodal age distribution has been noted, with peaks in young adults and individuals in their mid-fifties. The syndrome affects all races and ethnicities5.

PTS presents acutely with pain around the shoulder girdle, radiating toward the arm and hand. This pain, often described as sharp or throbbing, is usually severe enough to wake the patient from sleep and may last for several days to weeks before subsiding gradually6. Following the resolution of pain, patients experience varying degrees of motor weakness, muscular atrophy, or sensory disturbances, primarily affecting the proximal arm muscles7.
The exact pathophysiological mechanisms underlying PTS are still not fully understood. It is hypothesized to involve an immune-mediated inflammatory response that leads to damage within the brachial plexus. Some researchers propose that the syndrome may be triggered by viral infections, surgical procedures, vaccinations, or strenuous physical activity, which somehow precipitate an autoimmune response against peripheral nerve fibers.

Recent studies have explored the potential genetic predisposition to PTS, suggesting that certain hereditary traits may increase susceptibility to the syndrome. Familial cases of PTS have been documented, indicating a possible genetic component that could predispose individuals to nerve inflammation and damage.

Diagnosing PTS can be challenging due to its sudden onset and the variety of its symptoms, which can mimic other neurological conditions. Diagnosis is primarily clinical, supported by the patient's history and physical examination findings.

Electromyography (EMG) and nerve conduction studies can help assess the extent of nerve involvement and exclude other differential diagnoses. Recent advances in imaging, particularly MRI and ultrasound, have improved the ability to visualize inflammatory changes in the brachial plexus and exclude other structural causes of the symptoms.

The management of PTS is primarily supportive and symptomatic. Initial treatment typically involves pain management with NSAIDs, corticosteroids, or opioid analgesics. Once the acute pain subsides, physical therapy is crucial to prevent muscle atrophy and maintain range of motion. In some severe cases, surgical intervention may be considered.

The prognosis of PTS varies widely. While many patients experience significant spontaneous recovery, especially in muscle strength, within the first two years, a substantial proportion may have residual weakness, sensory abnormalities, or pain. Early intervention and rehabilitation are crucial to improving outcomes.

Raising awareness about PTS among healthcare providers is critical for early diagnosis and management. Educating patients who undergo surgeries involving the shoulder or those engaging in activities that could trigger PTS is also vital for prevention and early treatment.

Ongoing research is essential to unraveling the complex pathophysiology of PTS and developing more effective treatments. Current studies focus on the molecular and immunological aspects of the syndrome, aiming to identify specific biomarkers for early diagnosis and targeted therapy.

The objective of this review article on Parsonage-Turner Syndrome is to provide a comprehensive update on the multiple facets of the condition, encompassing its epidemiology, clinical presentation, pathophysiology, diagnostic approaches, and management strategies.

The review aims to elucidate the syndrome’s pathogenic mechanisms, examining current theories regarding its immunological and genetic causes. It also seeks to clarify the relationship between the clinical manifestations and the anatomical features observed in affected individuals.

However, the article intends to summarize and evaluate the latest diagnostic modalities that enhance the accuracy of detecting this condition. Management strategies will be thoroughly reviewed, emphasizing evidence-based treatments, and integrating multidisciplinary approaches to optimize patient outcomes.

This study also plans to highlight gaps in the current knowledge and suggest directions for future research, ensuring that the medical community remains abreast of the latest advancements and understands the complex nature of Parsonage-Turner Syndrome.

This comprehensive synthesis of current evidence and new insights aims to assist healthcare professionals in refining their diagnostic and therapeutic approaches to this challenging and often underrecognized condition.

2. Methods

The research methodology for this review was designed to investigate Parsonage-Turner Syndrome (PTS), focusing on its physiopathology, diagnosis, and therapy. Multiple reputable databases were utilized to ensure comprehensive coverage of relevant scientific and medical literature, including PubMed, Scopus, SciELO, Embase, and Web of Science.
which were recognized for their extensive collections of peer-reviewed publications. Google Scholar was employed to access gray literature, which often includes significant studies not available in standard academic journals. The primary objective was to understand the current knowledge regarding the incidence, underlying mechanisms, and treatment options for PTS. To achieve this, search parameters were carefully crafted using relevant keywords such as "brachial plexus neuritis," "Parsonage-Turner syndrome," "neuralgic amyotrophy," "physiopathology," "diagnosis," and "therapy." This strategic combination of search terms ensured the retrieval of studies directly pertinent to the research objectives. Inclusion criteria encompassed a broad spectrum of study designs, including randomized controlled trials, cohort studies, case-control studies, systematic reviews, and meta-analyses. This approach aimed to capture diverse evidence and perspectives on PTS. Exclusion criteria were established to filter out studies focusing on unrelated pathologies, non-PTS-related nerve conditions, or other age groups. Two independent reviewers initially screened each study's title and abstract for relevance and compliance with predefined criteria to ensure methodological rigor. Any discrepancies between the reviewers were resolved through consultation with a third reviewer, thereby minimizing bias and ensuring consistent selection. This dual-review process ensured that the final dataset comprised studies meeting the highest standards of relevance and quality. This systematic approach to the literature review provided a solid foundation for evaluating and synthesizing the findings. It ensured that this study's conclusions were based on a comprehensive and critically assessed body of scientific evidence regarding Parsonage-Turner Syndrome, including its genetic factors, diagnostic advancements, treatment protocols, and impact on the patient's quality of life.

3. Results and Discussion

The discussion surrounding Parsonage-Turner Syndrome (PTS) reflects a deepening understanding of this complex condition, yet significant gaps persist in areas ranging from genetic predisposition to effective treatment protocols.

Despite its clear clinical presentation, the specific genetic factors predisposing individuals to PTS remain elusive. While familial cases suggest a genetic component, the exact genes and their interactions have not been fully delineated. Current hypotheses propose a multifactorial origin involving genetic susceptibilities, such as variations in immune response genes, and environmental triggers like viral infections or immunizations.

Advanced genomic studies, including genome-wide association studies, could provide insights into the genetic architecture of PTS, potentially identifying risk alleles and gene-environment interactions that contribute to the syndrome.

Genetic researchers continue to explore the possibility of a polygenic nature or significant gene-environment interactions influencing PTS. Genomic technologies, such as whole-exome sequencing or genome-wide association studies, hold promise in identifying genetic variants predisposing individuals to PTS, which could lead to predictive genetic testing and more precise familial counseling.

Innovations in imaging technology play a pivotal role in diagnosing PTS. High-resolution magnetic resonance imaging (MRI) and ultrasound have substantially improved our ability to visualize nerve damage. The advent of magnetic resonance neurography (MRN) has further enhanced this capability by providing detailed images of peripheral nerve structures, potentially allowing for earlier and more accurate diagnoses. Future advancements may include the integration of artificial intelligence to interpret these complex images, offering even greater diagnostic precision.

Biomarkers have immense potential to transform PTS's diagnostic and therapeutic landscape. A comprehensive biomarker panel, including inflammatory markers and particular antibodies, could facilitate rapid, noninvasive diagnosis and monitor therapeutic responses, enabling timely interventions that improve outcomes.

Due to its unpredictable prognosis, treatment protocols for PTS vary, ranging from conservative approaches like physical therapy to aggressive interventions such as corticosteroids and immunoglobulin therapy.

The efficacy of these treatments remains variable, highlighting the need for standardized, evidence-based guidelines. However, exploring tailored immunomodulatory treatments and biological agents targeting specific inflammatory pathways may provide more effective treatment options.

The effectiveness of treatments for PTS varies, reflecting the syndrome's unpredictable nature and diverse manifestations. Current management strategies are primarily empirical, with corticosteroids and intravenous immunoglobulins (IVIG) frequently used despite limited high-quality evidence for their efficacy.
Physical therapy is a cornerstone of treatment, focusing on maintaining muscle function and range of motion. The optimal treatment protocols for PTS are yet to be established through rigorous, controlled clinical trials. Developing evidence-based guidelines will require a coordinated effort to study the long-term outcomes of various therapeutic approaches and their impact on disease progression.

The absence of a standardized treatment and diagnostic protocol for Parsonage-Turner Syndrome (PTS) is attributed to several complex factors. One of the primary challenges in standardizing treatment lies in the heterogeneity of PTS presentations. The disease's symptoms, severity, and progression vary significantly among individuals, complicating the development of a universally applicable treatment regimen. This variability makes it difficult to establish uniform treatment protocols and challenges the efficacy of potential therapies across different patient groups.

The diagnosis of PTS suffers from limitations due to the lack of specific biomarkers and the reliance on clinical symptoms and imaging findings for confirmation. The syndrome often presents with symptoms that mimic other neurological disorders, leading to potential misdiagnosis or delayed diagnosis. The diagnostic process is further complicated by the transient nature of some symptoms, which may resolve before the patient seeks medical advice.

Another significant hurdle is the unclear etiology of PTS. While genetic predispositions and autoimmune responses are suspected, the exact triggers and pathophysiological mechanisms remain poorly understood. This gap in understanding the fundamental causes of PTS hampers the development of targeted diagnostics and treatments.

The development of standardized diagnostic and treatment protocols for Parsonage-Turner Syndrome is impeded by the disease's variable clinical manifestations, the absence of specific diagnostic markers, unclear etiology, and the challenges posed by its rarity. These factors collectively hinder the advancement of universally accepted medical practices for managing PTS.

The link between PTS and various systemic health conditions, such as viral infections and autoimmune disorders, suggests a complex interplay that contributes to the syndrome's pathogenesis. These relationships could uncover new therapeutic targets and enhance our overall approach to treatment.

Long-term effects of PTS, including persistent pain, muscle atrophy, and sensory deficits, necessitate a multidisciplinary approach to management. This should include advanced physical therapy techniques, neuromuscular electrical stimulation, and potentially regenerative medicine approaches like stem cell therapy, aiming at functional recovery and improving quality of life.

Social and psychological impacts of PTS are profound. Patients often face significant challenges in daily activities and employment, which can lead to social isolation and psychological distress. Comprehensive management strategies should, therefore, also incorporate psychological support and social services to improve patients' overall well-being and societal integration.

The potential long-term impacts of PTS on patients' quality of life are significant and can include persistent pain, sensory disturbances, and functional disability due to muscle atrophy and weakness. These sequelae can profoundly affect personal and professional aspects of life, leading to psychological distress and socioeconomic challenges.

A comprehensive multidisciplinary approach is essential for addressing these impacts. To provide holistic care, this approach should integrate medical treatments with physical and occupational therapies, pain management, psychological support, and social services.

Various specialists such as neurologists, physiotherapists, occupational therapists, pain specialists, and psychologists, this strategy aims to enhance recovery, improve functional outcomes, and support patients in adapting to lifestyle changes imposed by the syndrome.
Considering the current treatment landscape, there is an ongoing debate about the optimal approach for active PTS, with choices often tailored to individual patient circumstances\(^4\). Research into the comparative effectiveness of different treatment modalities, especially the roles of immunotherapy versus corticosteroids, is crucial to optimizing patient care\(^5\).

Looking to the future, the research community remains hopeful about developing specific, targeted therapies based on a deeper understanding of PTPTS's underlying mechanisms\(^3\). Clinical trials and studies are expected to play a critical role in this endeavor, potentially leading to breakthroughs that could offer long-term relief and improve the lives of those affected by this debilitating condition\(^4,29\).

While our understanding of Parsonage-Turner Syndrome has advanced, significant research efforts are still required to fill the existing knowledge gaps\(^3\). Future studies should focus on genetic factors, advanced diagnostic tools, effective biomarkers, and comprehensive, evidence-based treatment protocols. These areas will enhance our understanding and significantly improve the quality of life for those affected by PTS\(^5,56\).

### 4. Conclusion

In conclusion, Parsonage-Turner Syndrome (PTS) remains poorly understood, with significant gaps in its genetic basis, diagnostic methods, and treatment protocols. Advances in genetic research, imaging, and biomarkers are promising but incomplete.

The variability in treatment efficacy underscores the need for standardized, evidence-based approaches. Future research should focus on identifying genetic markers, improving diagnostics, and developing targeted therapies to enhance patient outcomes. A multidisciplinary approach is crucial for comprehensive management of PTS, addressing its multifaceted impacts on patients' lives.

### Compliance with ethical standards

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**Disclosure of conflict of interest**

The authors declare that there is no conflict of interest.

### References


