THE PATTERN OF CLINICAL PRESENTATION
AND AETIOLOGY OF NON TRAUMATIC
QUADRIPLASESIS IN ADULT SUDANESE PATIENTS

2002 – 2003

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degree of Clinical MD, April 2004

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# Table of content

<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>Content</td>
<td>I</td>
</tr>
<tr>
<td>Dedication</td>
<td>III</td>
</tr>
<tr>
<td>Acknowledgement</td>
<td>IV</td>
</tr>
<tr>
<td>Abstract</td>
<td>V</td>
</tr>
<tr>
<td>Abbreviation</td>
<td>VIII</td>
</tr>
<tr>
<td>Introduction and literature review</td>
<td>1</td>
</tr>
<tr>
<td>Objectives</td>
<td>37</td>
</tr>
<tr>
<td>Patients &amp; Methods</td>
<td>38</td>
</tr>
<tr>
<td>Results</td>
<td>40</td>
</tr>
<tr>
<td>Discussion</td>
<td>66</td>
</tr>
<tr>
<td>Conclusion</td>
<td>73</td>
</tr>
<tr>
<td>Recommendations</td>
<td>74</td>
</tr>
<tr>
<td>References</td>
<td>75</td>
</tr>
<tr>
<td>Appendix</td>
<td></td>
</tr>
</tbody>
</table>
## List of Tables and Figures

<table>
<thead>
<tr>
<th>Table/Figure</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>Table-1</td>
<td>47</td>
</tr>
<tr>
<td>Table-2</td>
<td>48</td>
</tr>
<tr>
<td>Table-3</td>
<td>49</td>
</tr>
<tr>
<td>Table-4</td>
<td>50</td>
</tr>
<tr>
<td>Table-5</td>
<td>51</td>
</tr>
<tr>
<td>Table-6</td>
<td>52</td>
</tr>
<tr>
<td>Table-7</td>
<td>53</td>
</tr>
<tr>
<td>Figure-1</td>
<td>54</td>
</tr>
<tr>
<td>Figure-2</td>
<td>55</td>
</tr>
<tr>
<td>Figure-3</td>
<td>56</td>
</tr>
<tr>
<td>Figure-4</td>
<td>57</td>
</tr>
<tr>
<td>Figure-5</td>
<td>58</td>
</tr>
<tr>
<td>Figure-6</td>
<td>59</td>
</tr>
<tr>
<td>Figure-7</td>
<td>60</td>
</tr>
<tr>
<td>Figure-8</td>
<td>61</td>
</tr>
<tr>
<td>Figure-9</td>
<td>62</td>
</tr>
<tr>
<td>Figure-10</td>
<td>63</td>
</tr>
<tr>
<td>Figure-11</td>
<td>64</td>
</tr>
<tr>
<td>Figure-12</td>
<td>65</td>
</tr>
<tr>
<td>Figure-13</td>
<td>66</td>
</tr>
<tr>
<td>Figure-14</td>
<td>67</td>
</tr>
<tr>
<td>Figure-15</td>
<td>68</td>
</tr>
<tr>
<td>Figure-16</td>
<td>69</td>
</tr>
</tbody>
</table>
DEDICATION

TO

MY PARENTS
Acknowledgement

I would like to thank Dr. Abbashar and Dr. Ammar for their great support and invaluable efforts and advice during all the research steps.

My great thanks to Dr. Zeinab, Dr. Weam, Eng. Imad, Eng. Elrasheed and Mr. Mohammed for their great support.

All thanks to those patients who are willingly and generously included in the study, hoping to enjoy happy life and recovery.

Finally, my thanks to my family for continuous encouragement and supporting.
ABSTRACT

Background:

Quadriplegia is a major disabling health problem. Its importance is due to the incapacitating disability that often persists despite treatment. It encompasses a large range of disease entities ranging from degenerative, demyelination, infection, familial, autoimmune, cerebrovascular accident and space occupying lesion in the brain.

Aim of the study:

Is to study the etiologies and clinical presentation of quadriplegia in Sudanese patients.

Material and methods:

This is a descriptive cross-sectional study of 100 patients admitted in Al Shaab Teaching Hospital from Jan.2002-Oct.2003, presented with four limbs weakness.

Results:

There were 55 male and 45 female, more than half of the patients (55%) were in the age group of 18-44. Patients were originally from different states and different racial groups. 75% of the patients presented with gradual onset. Weakness was the presenting complaint in all patients. 39% of the patients were wheelchair bounded.
CSF analysis showed high protein in 12 patients (75%) with acute inflammatory demyelinating polyradiculoneuropathy, and in 14 patients (94%) with chronic inflammatory demyelinating polyradiculoneuropathy. Creatine phosphokinase was significantly high in all patients with muscular dystrophy. MRI detects the lesion in all patients with cervical cord compression.

**Conclusion:**
Cervical cord compression, acute inflammatory demyelinating polyradiculopathy, chronic inflammatory demyelinating polyradiculopathy, motor neuron disease and muscular dystrophy constituted the etiological bulk of quadriparesis in the study.
ملخص الأطروحة

يعتبر الشلل الرباعي من المعوقات الصحية الهامة وذلك يعود إلى العجز الذي يخلفه المرض احيانا رغم العلاج. وتتراوح أسباب الشلل الرباعي بين امراض انحلالية؛ زوال النخاعين؛ خمجية، وراثية، صدمة عصبية، وأورام دماغية.

الهدف من الدراسة هو معرفة مسببات الشلل الرباعي و مظاهرها السريرية. أجريت هذه الدراسة المقطعية الوصفية في مستشفى الشعب التعليمي في الفترة من يناير 2002 الى أكتوبر 2003 لمانعة مريض يعانون من ضعف كل الأطراف العليا والسفلى.

أظهرت النتائج أن 55% من المرضى كانوا من الذكور و 45% من الإناث. 55% من المرضى تتراوح أعمارهم بين 18-44 سنة و هم من مختلف أنحاء السودان و أعماره. 75% من المرضى ظهرت عليهم أعراض المرض بصورة تدريجية. كل المرضى كانوا يعانون من ضعف الأطراف الأربع. 39% من المرضى لا يستطيعون الحركة على أقدامهم اطلاقا.

فحص سائل النخاع الشوكي كان دائما لدى 12 مريض بداء اعتلال الإعصاب المتعددة زوال النخاعين الحاد؛ وكان أيضا دائما لدى 14 مريض بداء اعتلال الإعصاب المتعددة زوال النخاعين المزمن. فحص انزيم الكرباتيين فسفوكايناز كان ايجابيا و دائما لدى كل مرضى الحثل العضلي. التخطيط بالرنين المغناطيسي دعم تشخيص كل حالات انضغاط الحبل الشوكي الرقبي.

تشكل كل من انضغاط الحبل الشوكي الرقبي (26%); اعتلال الإعصاب المتعددة زوال النخاعين الحاد (16%); اعتلال الإعصاب المتعددة زوال النخاعين المزمن (15%); داء العصبون الحركي (10%); والحثل العضلي (10%) اسباب الاصحاب الخزل الرباعي بهذه الدراسة.
# ABBREVIATION

<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>AAFB</td>
<td>Alcohol Acid Fast Bacilli</td>
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<tr>
<td>AIDS</td>
<td>Acquired Immunodeficiency Disease</td>
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<tr>
<td>ARDS</td>
<td>Adult Respiratory Distress Syndrome</td>
</tr>
<tr>
<td>ATM</td>
<td>Acute Transverse Myelitis</td>
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<tr>
<td>CMT</td>
<td>Charcot-Marie-Tooth.</td>
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<tr>
<td>CNS</td>
<td>Central Nervous System</td>
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<tr>
<td>CPK</td>
<td>Creatine Phosphokinase</td>
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<tr>
<td>CSF</td>
<td>Cerebro Spinal Fluid</td>
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<td>CT</td>
<td>Computerized Tomography</td>
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<td>ECG</td>
<td>Electrocardiography</td>
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<tr>
<td>EEG</td>
<td>Electroencephalography</td>
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<tr>
<td>ELISA</td>
<td>Enzyme-Link Immuno-Sorbant Assay</td>
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<td>EMG</td>
<td>Electromyography</td>
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<td>GBS</td>
<td>Gillian- Barrie Syndrome</td>
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<td>GCS</td>
<td>Glasgow Coma Scale</td>
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<td>HIV</td>
<td>Human Immunodeficiency Virus</td>
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<tr>
<td>HLA</td>
<td>Human Leukocyte Antigen</td>
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<tr>
<td>I.C.U.</td>
<td>Intensive Care Unit</td>
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<td>LMNL</td>
<td>Lower Motor Neuron Lesion</td>
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<tr>
<td>MG</td>
<td>Myasthenia Gravis</td>
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<tr>
<td>MND</td>
<td>Motor Neuron Disease</td>
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<tr>
<td>MRA</td>
<td>Magnetic Resonance Angiography</td>
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<tr>
<td>MRI</td>
<td>Magnetic Resonance Image</td>
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<tr>
<td>MS</td>
<td>Multiple Sclerosis</td>
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<td>NC</td>
<td>Nerve Conduction Study</td>
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<td>SLE</td>
<td>Systemic Lupus Erythematosus</td>
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<tr>
<td>MNL</td>
<td>Upper Motor Neuron Lesion</td>
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<tr>
<td>WBC</td>
<td>White Blood Cell Count</td>
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<td>WHO</td>
<td>World Health Organization</td>
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**INTRODUCTION:**

Quadriparesis is weakness affecting all four limbs, can result from upper motor neuron disease (localized to the upper cervical spinal cord or above), diffuse lower motor neuron disease, or myopathy (1).

**Acute Quadriparesis:**

The differential diagnosis for acute quadriparesis with onset over minutes includes many diseases of upper motor neuron e.g. (anoxia, hypotension, brainstem or cervical cord ischemia, trauma and systemic metabolic abnormalities) and also rare forms of myopathic weakness (systemic toxins or periodic paralysis) (1).

Onset over hours to a few days may be due to upper motor neuron, lower motor neuron or myopathic disorders (1).

All three pattern of weakness are usually associated initially with hypotonia (1).

If acute quadriparesis is associated with stupor or coma, the evaluation begins with a CT-Scan of the brain (1).

If upper motor neuron signs are present but the patient is alert, the initial test is usually MRI of the cervical cord (1).

If weakness is lower motor neuron, myopathic, or uncertain in origin, the clinical approach starts with an electro diagnostic study (1).

Acute demyelinating polyneuropathy is an important diagnostic possibility in this setting (1).

**Sub acute or chronic quadriparesis:**

When quadriparesis due to upper motor neuron disease develops over weeks, months, or years, the distinction between disorders of the cerebral hemispheres, brainstem, and cervical spinal cord is usually possible by clinical criteria alone (1).

The diagnostic approach begins with MRI of the clinically suspected site (1).
Lower motor neuron disease present with weakness that is most profound distally where as myopathic weakness is typically proximal.
The evaluation then begins with EMG and nerve conduction studies\(^{(1)}\).

**NEUROANATOMY OF THE MOTOR SYSTEM:**

There are three systems:

- **The corticospinal or (pyramidal) system:** originate in the cerebral cortex and delivers information to the anterior horn cells of the spinal cord. This system enables purposive, skilled, strong and organized movement to take place. Defective function within the pathway is recognized by a distinct pattern of loss of skilled voluntary movement, spasticity and reflex changes\(^{(2)}\).

- **The extra pyramidal system:** facilitate fast, fluid movement that the corticospinal system has generated. Defective function is recognized usually by slowness (bradykinesia), stiffness (rigidity) and/or disorders of movement (rest tremor, chorea and dyskinesias)\(^{(2)}\).

- **The cerebellum and its connections:** have a role in coordinating the smooth movement initiated by the corticospinal system, and in the regulation of balance\(^{(2)}\).

**The corticospinal or (pyramidal) system:**

The corticospinal tracts originate in the neurons of the fifth layer of the cortex and terminate at the motor nuclei of the cranial nerves and anterior horn cells of the spinal cord. The nerve fiber pathways of a particular importance in clinical diagnosis congregate in the internal capsule and cross in the medulla (decussating of the pyramids). Passing to the contra lateral halves of the spinal cord as the crossed lateral corticospinal tracts\(^{(2)}\).
This is the pyramidal system disease of which causes upper motor neuron (UMN) lesions.

A small proportion of the corticospinal outflow remains uncrossed (the anterior corticospinal tracts) but this is not of relevance in clinical practice.

The lower motor neuron (LMN) is the motor pathway from the anterior horn cell (or cranial nerve nucleus) via a peripheral nerve to the motor end plate (2).

The motor unit consists of the a single anterior horn cell, the single fast-conducting motor nerve fiber that leaves the spinal cord via the anterior root, and the group of muscle fibers (100-200) being supplied via the mixed peripheral nerve. Anterior horn cell activity is modulated by impulses from the corticospinal tracts, the extra pyramidal system, the cerebellum and afferent fibers from the posterior roots (2).

CERVICAL CORD COMPRESSION

Cervical Spondylosis:

Cervical spondylosis results from chronic cervical disc degeneration, with herniation of disc material, secondary calcification and associated with osteophytic out growth. One or more of the cervical nerve roots may be compressed, stretched, or angulated, and myelopathy may also develop as a result of compression, vascular insufficiency, or recurrent minor trauma to the cord (3).

Patients present with neck pain and restricted head movement, occipital headache, radicular pain and other sensory disturbances in the arms, weakness of the arms or legs, or some combination of these symptoms (3).

Examination generally reveals that lateral flexion and rotation of the neck are limited. A segmental pattern of weakness or dermatomal sensory loss or both may be found unilaterally or bilaterally in the upper limbs, and tendon reflexes mediated by the affected root or roots are depressed (3).
The C5 and C6 nerve roots are most commonly involved, and the examination frequently then reveals weakness of the muscle supplied by these roots (deltoid, supra. and infraspinatus, biceps, brachioradialis), pain or sensory loss about the shoulder and outer border of the arm and forearm, and depressed biceps and brachioradialis reflexes. Spastic paraparesis may also be present if there is an associated myelopathy, sometimes accompanied by posterior column or spinothalamic sensory deficit in the legs³.

Degeneration of cervical discs and joints may occur in adolescents but is more common after age of 40. Degeneration is progressive and is marked by gradual narrowing of the disc space, as demonstrated by x-ray. Osteocartilaginous proliferation occurs around the margin of the vertebral body and gives rise to osteophytic ridges that may encroach upon the intervertebral foramina and spinal canal, causing compression of the neurovascular contents⁴.

Osteoarthritis of the cervical spine is often a symptomatic but may cause diffuse neck pain, radicular pain or myelopathy. Myelopathy develops insidiously and is manifested by numb, clumsy hands. Some patients also complain of unsteady walking, urinary frequency and urgency, or electrical shock sensation with neck flexion or extension (Lhermitte’s sign). Weakness, sensory loss and spasticity with exaggerated reflexes develop below the level of the spinal cord compression⁴.

Plain radiographs of the cervical spines show osteophyte formation, narrowing of the disc spaces and encroachment on the vertebral foramina, but many changes are common in middle-aged persons and may be unrelated to the presenting complaint⁴.

CT or MRI helps to confirm the diagnosis and exclude other structural causes of the myelopathy⁴.

Restriction of neck movements by a cervical collar may relieve the pain. Operative treatment may be necessary to prevent further progression if there is a
significant neurological deficit or if root pain is severe, persistent, and unresponsive to conservative measures\(^{(3)}\).

Atlantoaxial subluxation may occur in patient with rheumatoid arthritis, regardless of the severity of the disease. Inflammation of the synovial structures resulting from erosion and laxity of the transverse ligament can lead to neurological signs of the spinal cord compression\(^{(4)}\).

Treatment may vary with from use of a cervical collar or more rigid bracing to operative treatment, depending on the degree of subluxation and neurologic progression. Surgical treatment may involve stabilization of the cervical spine\(^{(4)}\).

**Spontaneous Cervical Spinal Epidural Hematoma:**

It is an uncommon entity. In the absence of trauma spinal epidural hematoma are considered spontaneous\(^{(5)}\).

The most common causes are anticoagulant therapy, vascular anomalies, hypertension, and blood spinal surgery\(^{(6)}\).

In up to 40\% of cases, no underlying case can be identified\(^{(7)}\).

The clinical picture is that of acute spinal cord compression, beginning with local back and radicular pain followed by sensory changes and finally motor weakness or paralysis. This symptom complex evolves within hours\(^{(8)}\).

The standard therapy has been prompt evacuation of the haematoma, usually with good neurological recovery\(^{(9)}\).

Spontaneous resolution and complete recovery without surgical decompression was reported in few cases\(^{(10)}\).

**Spinal Cord Tumors:**

10\% of the spinal cord tumors are inframedullary. Ependymoma is the most common type of inframedullary tumor. Extramedullary tumors may be extradural or
intradural in location. The primary extramedullary tumors, neurofibromas and meningiomas, are relatively common; are benign and may be intra or extradural\textsuperscript{(11)}.

Carcinomatous metastasis, lymphomatosis or leukemic deposit and myeloma are usually extradural, in the case of metastases; the prostate, breast, lungs and kidney are common primary sites\textsuperscript{(11)}.

Tumors may lead to spinal cord dysfunction by direct compression, by ischemia secondary to arterial or venous obstruction and, in cases of intramedullary lesion, by invasive infiltration\textsuperscript{(11)}.

CT myelography or MRI may be necessary to identify and localize the site of cord compression\textsuperscript{(11)}.

CSF is often xanthochromic and contains high protein concentration, with normal cells count and glucose concentration\textsuperscript{(11)}.

Treatment with high dose of dexamethasone is used to reduce cord swelling and irradiation therapy to delay the onset of disability. Intramedullary tumors are treated by surgical excision and decompression\textsuperscript{(11)}.

\textit{ACUTE INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY:}

\textbf{Background:} Acute inflammatory demyelinating polyradiculoneuropathy (AIDP) is an autoimmune process that is characterized by progressive weakness many variants exist. This associated with distal paraesthesia and loss of deep tendon reflexes\textsuperscript{(12)}.

Myelin breakdown and axonal degeneration were observed in nerve biopsies from patients with (AIDP) by Haymaker and Kernohan in 1949. An allergic etiology was suggested by Krucke in 1955 after he observed lymphocytic infiltrates within biopsy specimens\textsuperscript{(13)}.
Pathophysiology: AIDP is believed to be caused by an immunologic attack that is directed against myelin components. This results in a demyelinating Polyneuropathy, both cellular and humoral immune mechanisms appear to play a role. Early inflammatory lesions consist of a lymphocytic infiltrate that is adjacent to segmental demyelination. Macrophages are more prominent several days later\(^\text{13}\).

The changes are observed in nerve roots, and cranial nerves. In acute motor axonal neuropathy (AMAN, an AIDP variant), deposited complement is found at the nodes of Ranvier. While myelin often is left undamaged\(^\text{14}\).

Damage to the myelin sheath leads to segmental demyelination. This results in decreased nerve conduction velocity and, at times, conduction block. Occasionally, in more severe and rapid cases, axonal degeneration also is observed which results in wallerian degeneration\(^\text{14}\).

Causes: AIDP is thought to be caused by a dysregulated immune response against myelin. This response may be triggered by several illnesses and conditions. Two-thirds of patients with AIDP recall an antecedent upper respiratory or gastrointestinal infection, e.g., campylobacter jejuni\(^\text{15}\).

Several cases have been reported after immunization against rabies, influenza, measles, mumps, or rubella\(^\text{15}\).

History: Classically, AIDP present as an ascending paralysis. Even in these cases, the clinical presentation and course vary. Additionally, many variants exist that differ markedly from classic AIDP in disease onset or course.

The hallmark of classic AIDP is progressive weakness that usually begins in the feet before involving all four limbs. At presentation, 60% of patients have weakness in all four limbs. Weakness plateaus at two weeks after onset in 50% of
patients and by four weeks in over 90%. It is usually symmetric, although mild asymmetry is not uncommon early in the disease course. In the arms, weakness may be worse proximally than distally. At presentation, half of patients have some facial weakness, although only 5% have varying degrees of ophthalmoplegia.\(^{(12,16)}\)

Forty percent have oropharyngeal or respiratory weakness at presentation. Improvement in strength usually begins 1-4 weeks after the plateau. About one third of patients require mechanical ventilation because of respiratory failure.\(^{(12)}\)

Sensory system, mild to moderate severe paraesthesias in the distal limbs are common and often precede the onset of weakness by 1 or more days. Proximal sensory changes are uncommon but may occur in more severe cases of AIDP.\(^{(17)}\)

About two third of patients have one or more autonomic abnormalities. Sustained sinus tachycardia is the most common dysfunction. Postural hypotension leading to presyncope or syncope can occur. Sweating dysfunction is common but rarely noted by patients. Urinary retention and constipation are more likely to occur late in the course of AIDP.\(^{(18)}\)

The Miller–Fisher variant is presented with ophthalmoplegia, areflexia and ataxia.\(^{(19)}\)

**Physical:** Although patients often report only weakness in the leg, careful examination usually demonstrates arm weakness (proximally and distally some patients with Miller–fisher or other regional variants may have weakness of cranial muscles only).\(^{(12)}\)

Deep tendon reflexes, hyporeflexia or areflexia is seen in 70% of patients at presentation and eventually in all patients. A progressive decrease in reflexes is a useful finding that may precede EMG changes.\(^{(12)}\)

Fluctuations in heart rate, specifically a sustained sinus tachycardia, are seen often. Some intubated patients also may have bradycardia, especially after vagal stimulation with Valsalva and/or tracheal suctioning maneuvers.\(^{(18)}\)
Orthostatic hypotension can occur and is likely due to dysfunction of baroreceptor reflex\(^{(18)}\).

At times, the labile blood pressure is observed with severe hypertension that may be due to dysfunction of the afferent of the baroreceptor reflex.\(^{(18)}\).

**C.S.F:** Increased CSF protein without an increased WBC count (albuminocytologic dissociation) is observed classically in AIDP. However, this finding is not specific to AIDP. About two thirds of patients have this CSF finding during the first week of symptoms and 82% have it by 2 weeks after symptoms onset. Although protein values can be elevated by 10–fold or more, no association exists between protein level and clinical severity\(^{(14)}\).

Nerve studies (NCS) can document demyelination, the hallmark of AIDP. Early on findings of NCS studies are often normal. However, 90% are abnormal within 3 weeks of symptom onset\(^{(20)}\).

Pulmonary function tests, useful in determining the timing of unit (ICU) transfer and intubation, should be performed in all patients\(^{(21)}\).

Transfer to ICU generally is indicated when Forced vital capacity (FVC) is less than 20 ml/kg. Intubation usually is warranted when FVC drop to 15 ml/kg or negative inspiratory pressure drop to less than –25 cm H\(_2\)O\(^{(21)}\).

Electrocardiogram (ECG) and cardiac monitoring can be helpful when arrhythmias occur. Other possible abnormalities include atrioventricular block, QRS widening, and T-wave abnormalities\(^{(21)}\).

**Medical care:** Mechanical ventilator assistance is required in about one third of patients with AIDP and lasts for an average of 49 days. Tracheostomy usually is recommended if mechanical ventilation will be required for more than 2-3 weeks\(^{(21)}\).
Cardiac monitoring is necessary. Chronic sinus tachycardia often responds to beta–blockers or calcium channel blockers. Bradycardia requires atropine treatment, if symptomatic. Heart block may require temporary pacing. Hypertension responds well to beta–blockers \(^{(21)}\).

Immunomodulation with IV immunoglobulin and plasmapheresis has led to faster recovery, relatively mild disability, and shorter hospital stays \(^{(22)}\).

**Prognosis:** About 75% of patients have an excellent recovery and regain their pre-morbid condition. Some of these patients experience easy fatigability for many years. Death occurs in only 2-6% of patients and is usually due to cardiac arrest, ARDS, pulmonary embolism, severe bronchospasm, pneumonia, and sepsis \(^{(23)}\).

**CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY:**

**CIDP** is an acquired immunological mediated disorder, is clinically similar to GBS except in that it has relapsing or steadily progressive course over months or years \(^{(24)}\).

In the relapsing form, partial recovery may occur after some relapses, but in other instances there is no recovery between exacerbations \(^{(24)}\). Although remission may occur spontaneously with time, the disorder frequently follows a progressive downhill course leading to severe functional disability \(^{(24)}\).

CSF shows elevated protein level not accompanied by pleocytosis and electrophysiological changes \(^{(25)}\).

Electro diagnostic studies show marked slowing in motor and sensory conduction, and focal conduction block. Signs of partial denervation may be present.
owing to secondary axonal degeneration. Nerve biopsy may show chronic perivascular inflammatory infiltrates, without accompanying evidence of vasculitis \(^{(24)}\).

Corticosteroids may be effective in arresting or reversing the downhill course \(^{(24)}\).

Treatment is usually begun with prednisolone 60mg daily, for 2-3 months or until a definite response has occurred. If no response has occurred despite 3 months of treatment, a higher dose may be tried. In responsive cases, the dose is gradually tapered, but most of patients become corticosteroids-dependent, often requiring prednisolone 20mg daily on alternate days, on along term basis \(^{(24)}\).

Patients unresponsive to corticosteroids may benefit instead from treatment with cytotoxic drugs such as azathioprine, cyclophosphamide or cyclosporine and administration of high dose intravenous, immunoglobulin treatment (e.g:400mg/kg/day) produce clinical improvement lasting for weeks to months \(^{(24)}\).

* MUSCULAR DYSTROPHY:

These inherited myopathic disorders are characterized by progressive muscle weakness and wasting \(^{(26)}\).

They are subdivided by mode of inheritance, age of the onset and the clinical features \(^{(26)}\).

In the Duchenne type, pseudo hypertrophy of muscles frequently occurs at some stage; intellectual retardation is common; and there may be skeletal deformities, muscle contractures and cardiac involvement. The serum CPK level is increased, especially in Duchenne and Becker varieties, and mildly increased in the limb girdle dystrophy. EMG may help to confirm that the weakness is myopathic rather than neurogenic \(^{(26)}\).
A genetic defect on the short arm of the X chromosome has been identified in Duchen-ne dystrophy. The affected gene codes for the protein dystrophin, which is markedly reduced or absent from the muscle of patients with the disease (26).

Dystrophin level is generally normal in the Becker variety, but the protein is qualitatively altered.

It is important to encourage patients to lead as normal live as possible. Prolonged bed rest must be avoided, inactivity often lead to worsening of underlying muscle disease (26).

Physical therapy and orthopedic procedures may help to counteract deformities or contractures (26).

**Limb girdle muscular dystrophy:**

*Clinical features:* Muscle weakness affects both male and females, with onset ranging from late in the first decade to the fourth decade. Most limb-girdle muscular dystrophies are progressive and affect the pelvis and shoulder girdle muscles (27).

Respiratory insufficiency from diaphragm weakness may occur. The distribution of weakness and the rate of progression vary from family to another. Cardiac involvement results in congestive heart failure or arrhythmias; occasionally patients present with cardiomyopathy. Intellectual function remains normal (27).

*Laboratory features:* An elevated serum CPK level, myopathic EMG findings, and muscle biopsy features indicative of myopathy represent the characteristic changes in limb–girdle dystrophy (27).

*Treatment:* Supportive care, no specific treatment, wheel-chair may be essential or may be used to help preserve energy in work. Cardiac or respiratory muscle involvement may require individualized treatment (28).

**MOTOR NEURON DISEASE (MND):**
MND is characterized clinically by weakness and variable wasting of the affected muscles, without accompanying sensory changes (29).

MND mainly affect adults between 30 and 60 years of age. There is degeneration of the anterior horn cells in the spinal cord, the motor nuclei of the lower cranial nerves, and the corticospinal and corticobulbar tract. Generally have the poorest prognosis (29).

Classification: Five varieties have been distinguished on clinical grounds:

(A) Progressive bulbar palsy: Bulbar involvement predominates owing to disease processes affecting primarily the motor nuclei of the cranial nerves.

(B) Pseudo bulbar palsy: Bulbar involvement predominates in this variety also, but it is due to bilateral corticobulbar disease and thus reflects upper motor neuron dysfunction.

(C) Progressive spinal muscular atrophy: This characterized primarily by a lower motor neuron deficit in the limbs due to generation of the anterior horn cells in the spinal cord.

(D) Primary lateral sclerosis: There is a purely upper motor neuron deficit in the limbs (29).

(E) Amyotrophic lateral sclerosis (ALS): This is progressive, fatal motor neuron disease of unknown origin causes selective degeneration of both upper and lower motor neurons. Widespread loss of these neuronal links between the brain and skeletal muscle results in generalized muscle weakness and atrophy (30). Progressive respiratory insufficiency is usually the cause of death (30).

Onset of ALS is usually from middle to late adulthood, but persons of any age can be affected. Most cases of ALS occur sporadically, but an autosomal dominant is seen in 5% of patients (30).
Symptoms & Signs: The common clinical manifestation related to degeneration of upper and lower motor neuron function in ALS include: spasticity, hyperreflexia, muscle weakness, muscle atrophy, fasciculation, hyporeflexia, hypotonia, muscle cramps, dysartheria, dysphagia, exertional dyspnoea, respiratory failure, fatigue, insomnia, weight loss, tendon shortening and joint contracture\(^{(31)}\).

The disease begins with focal motor weakness at a specific brain stem or spinal level, then progress to involvement of contiguous spinal levels\(^{(31)}\).

The clinical hallmark of the ALS is the presence of widespread, purely motor signs of both upper and lower motor neuron dysfunction. ALS spares cognitive function, but this feature has potentially negative effects because patients remain fully aware of the progression of their disease\(^{(32)}\).

Diagnosis by EMG is confirmed by the presence of widespread, purely motor denervation of lower motor neurons\(^{(32)}\).

MRI studies are useful to exclude other possible disorders. Because ALS is a selective motor neuron disease, clear evidence of cognitive, visual, bladder, bowel, or sensory symptoms does not support the diagnosis\(^{(33)}\).

Treatment by Riluzole is an important break through because it prolongs survival by about 3 months, making it the only effective disease-specific therapeutic option\(^{(34)}\).

SPACE OCCUPYING LESIONS (SOL):

Primary intracranial tumors lead to focal deficit depending on their location\(^{(35)}\).

Meningiomas:

Meningiomas are derived from cells of the arachnoid granulations. These usually benign tumors, they attached to the dura and may invade the skull but almost never invade the brain\(^{(36)}\).
They frequently occur along the sagittal sinus and over the cerebral convexities, in the cerebellar-pontine angle and along the dorsum of the spinal cord. They are more frequent in women than men, with a peak incidence in middle age. Meningioma may be found incidentally on a CT or MRI scan, or may present with a focal seizure, a slowly progressive focal deficit, or symptoms of increased intracranial pressure. The radiologic image of a dural-based, extra-axial mass with dense, uniform contrast enhancement is essentially diagnostic, although a dural metastasis must also be considered\(^{(36)}\).

Meningioma compresses rather than invades adjacent neural structures. Increasingly, common with advancing age. Tumor size varies greatly. Symptoms vary with tumor site; unilateral exophthalmus (sphenoidal ridge), anosmia and optic nerve compression (olfactory groove)\(^{(36)}\).

Meningioma may lead to calcification and bone erosion, visible on plain x-rays of skull\(^{(36)}\).

Total surgical resection of benign meningioma is curative. If a total resection can not be achieved, local radiotherapy reduces the recurrence rate to less than 10\% \(^{(37)}\).

**Tuberculous meningitis & tuberculoma:**

Tuberculosis of the CNS accounts for about 5\% of extra pulmonary cases. It is seen most often in young children but also develop in adults, especially those who are infected with HIV. Tuberculous meningitis results from the haematogenous spread of primary or post primary pulmonary disease or from the rupture of a subependymal tubercle into the subarachnoid space. In more than half of the cases, evidence of old pulmonary lesion or miliary pattern is found on chest radiography. The disease may present as headache and mental changes or acutely as confusion, lethargy, altered Sensation and neck rigidity\(^{(38)}\).
Typically the disease evolves over 1 or 2 weeks- a course longer than that of bacterial meningitis. Paresis of the cranial nerves (ocular nerves in particular) is frequent finding, and the involvement of cerebral arteries may produce focal ischemia (38).

Hydrocephalus is common. Lumber puncture is the cornerstone of diagnosis. In general examination of the CSF reveals a high leucocytes count (usually with predominance of lymphocyte but often a predominance of neutrophil in the early stage), protein content of 1 to 8 g/l or (100-800 mg/dl) and low glucose concentration, however; any of these three parameters can be within the normal range .AAFB is seen on direct smear of CSF sediment in only 20% of cases. Imaging studies (CT and MRI) may show hydrocephalus and abnormal enhancement of basal cisterns or ependyma. If unrecognized tuberculous meningitis is uniformly fatal (38). This disease responds to chemotherapy; neurological sequelae are documented in 25% of treated cases; in most of which the diagnosis has been delayed. Glucocorticoids are useful adjunct to chemotherapy, especially in cases with cerebral edema or high CSF protein level (38).

**Tuberculoma:**

An uncommon manifestation of tuberculosis, presents as one or more space-occupying lesions and usually causes seizures and focal signs. CT or MRI reveals contrast-enhanced ring lesions, but biopsy is necessary to establish the diagnosis (38).

**Pott’s disease of cervical spine:**

Tuberculous arachnoiditis may arise at any level of the spinal cord in association with breakdown of a focus within the cord or meninges, or by extension from an adjacent area of in-apparent spondylitis .Cervical spine tuberculosis is a less common presentation but is serious (38).
POLYMYOSITIS – DERMATOMYOSITIS

**Background:** Polymyositis is a systemic disorder of unknown cause whose principal manifestation is muscle weakness. It is the most frequent primary myopathy in adults. When skin manifestations are associated with it, the entity is designated dermatomyositis. The true incidence is not known, since milder cases are frequently not diagnosed. The disease may affect person of any age group, but the peak incidence is in the fifth and sixth decades of life. Women are affected twice as commonly as men (39).

There is an increased risk of malignancy, especially in patients with dermatomyositis. The malignancy maybe evident initially or may not-become evident for months after the muscle disease presents. Ovarian cancer appears to be especially common, found in some series to affect more than 20% of women aged over 40 with dermayomyositis (40).

**Symptoms & sign:** Polymyositis may begin abruptly, although often it is gradual and progressive. The characteristic rash is dusky red and may be seen over the butterfly area of the face, neck, shoulder, and upper chest and back. Periorbital edema and purplish (heliotrope) suffusion over the upper eyelids are typical signs. Subungual erythema, cuticular telangiectases and scaly patches over the dorsum of the proximal interphalangeal and metacarpophalangeal joints (Gottron’s sign) are highly suggestive (39).

Muscle weakness chiefly involves proximal groups, especially of the extremities. Leg weakness (e.g.; difficulty rising from a chair or climbing steps) precedes arm symptoms. Neck flexor weakness occurs in 2/3 of the cases. Pain and
tenderness of affected muscles occur in 1/4 of the cases, and Raynaud’s phenomenon and joints symptoms may be associated. Atrophy and contractures occur late. Associated myocarditis is uncommon. Interstitial pulmonary disease, usually mild, is sometimes associated, and calcinosis may be observed, especially in children. Polymyositis may occur in association with sjogren's syndrome, SLE or scleroderma (39).

**Laboratory findings:** Measurement of serum level of muscle enzymes, especially CPK and aldolase, is the most useful in diagnosis and assessment of the disease activity (39).

The search for an occult malignancy should be routine for the patients (40).

Biopsy of clinically involved muscle, usually proximal, is the only specific diagnostic study (39).

**Management:** Most patients respond to corticosteroids. Often a daily dose of 40-60 mg or more of prednisolone is required initially and then reduced gradually (39).

**BRUCELLOSIS:**

Brucellosis is a zoonotic disease caused by several brucella species. The most common causes of human disease are brucella melitensis and brucella Suis (41).

Physical examination findings can include generalized lymphadenopathy and hepatosplenomegaly. Granulomas can be seen in the liver, spleen, bone marrow, lymph node, brain, skin and kidneys. Laboratory studies may show mild leucopenia or thrombocytopenia. Death occurs in less than 1% of cases, but brucellosis can cause significant illness for months to years (42).

**Neurobrucellosis:**
Is uncommon but serious and include meningoencephalitis, multiple cerebral or cerebellar abscesses, rupture mycotic aneurysms, myelitis, Guillain-Barre syndrome, cranial nerve lesions, hemiplegia, sciatica, myositis and rhabdomyolysis. Papillitis, papilloedema, retrobulbar-neuritis, optic atrophy and ophthalmoplegia due to lesions in cranial nerves III, IV and VI may occur in brucella meningoencephalitis.\(^{(43)}\).

Cerebrospinal fluid (CSF) pressure is usually elevated; the fluid may appear clear, turbid or hemorrhagic, the protein concentration and cell count (predominantly lymphocyte) are elevated; and the glucose concentration may be either reduced or normal.\(^{(43)}\).

In brucella meningitis, the organism may be cultured from the CSF.\(^{(43)}\).

**Treatment:** Antimicrobial agents include tetracycline, doxycycline, streptomycin, gentamycin, trimethoprim-sulfamethoxazole and quinolones.\(^{(44)}\)

Because relapse is common with monotherapy, combination therapies with doxycycline and rifampin (Rifadin) or with doxycycline and streptomycin have been recommended by WHO.\(^{(45)}\)

**STROKE:**

**Cerebral Infarction:**

Thrombosis or embolic occlusion of a major vessel leads to cerebral infarction. Causes include the disorders predisposing to transient ischemic attacks and atherosclerosis of the cerebral arteries. The resulting deficits depend upon the particular vessel involved and the extent of any collateral circulation. Cerebral ischemia leads to release of excitatory and other neuropeptides that may augment calcium flux into neurons, thereby leading to cell death and increasing the neurological deficit.\(^{(46)}\)
**Symptoms & signs:** Onset is usually abrupt, and there may be very little progression except that due to brain swelling. Clinical evaluation always includes examination of the heart and auscultation over the subclavian and carotid vessels to determine whether there are any bruits\(^{(47)}\).

**Imaging:** Radiography of the chest may reveal cardiomegaly or valvular calcification; the presence of a neoplasm would suggest that the neurological deficit is due to metastasis rather than stroke\(^{(46)}\). CT scan of the head (without contrast) is important in excluding cerebral hemorrhage \(^{(46)}\).

**Laboratory & Other Studies:** Investigations should include a complete blood count, erythrocytes sedimentation rate, blood glucose determination and serological tests for syphilis. Antiphospholipid antibodies promote thrombosis and are associated with an increased incidence of stroke. Similarly, elevated serum cholesterol and lipids may indicate an increased risk of thrombotic arrhythmia or recent myocardial infarction \(^{(47)}\).

**Treatment:** Early management of completed stroke consist of attention to general supportive measures during the acute stage, there may be marked brain swelling and edema, with symptoms and signs of increasing intracranial pressure and increasing neurological deficit or herniation syndrome\(^{(48)}\).

Attempt to lower the blood pressure of hypertensive patients during the acute phase of stroke should be avoided. Since there is loss of cerebral autoregulation, the lowering of the blood pressure may further compromise ischemic areas\(^{(48)}\).

Physical therapy has an important role in the management of patients with impaired motor function. Passive movements at an early stage will help in preventing contractures \(^{(48)}\).
In all cases, early mobilization and active rehabilitation are important. Occupational therapy may improve morale and motor skills, while speech therapy may be beneficial in patients with expressive dysphasia or dysarthria\(^{(48)}\).

**Prognosis:** Loss of consciousness after cerebral infarcts implies a poorer prognosis. Patients who have had a cerebral infarct are at risk for further strokes and for myocardial infarcts\(^{(47)}\).

**ENCEPHALITIS:**

Encephalitis is an acute inflammatory process that affects brain tissue and is most commonly caused by viral infection. Encephalitis resulting from viral infection manifests as either acute viral encephalitis or post infectious encephalomyelitis\(^{(49)}\).

The most frequently reported causes are; enteroviruses, herpes simplex virus type-1(HSV-1) and type-2 (HSV-2), and arthropod-borne viruses (Arboviruses). Before widespread of live measles, mumps, and rubella virus vaccine (MMR 11), mumps virus was also a common source of aseptic meningitis and encephalitis\(^{(50)}\).

Most patients with enterovirus CNS infection have mild generalized disease that resolves without ale. However, focal - infections and severe, even fatal, cases have been reported\(^{(51, 52)}\).

**Symptoms and Signs:** Include fever, headache, and behavioral changes. Subsequently, focal seizures, hemi paresis, cranial nerve deficits, and visual field abnormalities may occur\(^{(53)}\).

A score of less than 6 in GCS, age greater than 30 years, and presence of encephalitis for more than 4 days before initiation of therapy are predictive of severe sequelae or death. Most survivors of HSV encephalitis have neurological sequel

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ranging from impaired new learning to severe motor and sensory deficit, aphasia, and amnestic syndrome. Encephalitis may recur despite appropriate antiviral therapy (53).

Arboviruses are RNA viruses transmitted to vertebrate hosts by hematophagous arthropods, such as mosquitoes and ticks. Such viruses cause about 10% of all reported cases of sporadic encephalitis but may account for up to 50% of all cases in epidemic years (54).

HIV may be the primary cause of encephalitis in persons with AIDS; however, opportunistic infections also must be considered (55).

**Post-infectious Encephalomyelitis:**

Post infectious encephalomyelitis is characterized by periventricular inflammation and demyelination of brain tissue. In this disorder, peripheral blood lymphocytes react against myelin basic protein. Before widespread vaccination, Post infectious encephalomyelitis most commonly occurred after smallpox and measles infections. Recent years, the disease had been associated with various viral and bacterial infections (56, 57).

Patients may have a history of nonspecific respiratory or gastrointestinal illness for 1 to 2 weeks before the onset of neurological symptoms (58).

Acute cerebellar ataxia is a form of acute Post infectious encephalomyelitis following varicella infection.

Examination of the CSF in a patient with viral CNS infection reveals a normal or elevated opening pressure. A moderate elevation in white cells count is typical. Polymorph nuclear cells may predominate early in the illness, but are replaced by mononuclear cells within hours. Other typical CSF findings include a normal or elevated protein level and a normal glucose level. A routine Gram-stain should be obtained; in addition, CSF should be cultured for bacteria, viruses, fungi and mycobacterium (58).
Polymerase Chain Reaction (PCR) is being more widely used to detect viral DNA or RNA in CSF. This method has been reported to have greater than 95% sensitivity and 100% specificity for HSV DNA in patients with biopsy-proven HSV encephalitis\(^{(59)}\).

PCR techniques show excellent specificity and sensitivity in the diagnosis of enteroviral meningitis appears promising\(^{(51)}\).

EEG is a useful complementary test for diagnosis of encephalitis in patients with HSV encephalitis, EEG may reveal focal unilateral or bilateral periodic discharges localized in the temporal lobe\(^{(60)}\).

Brain biopsy is the definite test for diagnosis of acute encephalitis. Biopsy should be considered in cases in which the diagnosis is uncertain and response to therapy is poor\(^{(60)}\).

**Treatment:** Empirical therapy with intravenous acyclovir should be initiated when HSV encephalitis is suspected. The recommended dosage is 30 mg/kg/day in three divided doses for 14 to 21 days. Relapse may occur, and re-treatment should be considered.

Arboviral and enteroviral encephalitis is treated with symptomatic and supportive care. Patients with headache should rest in quiet, dark environment and take analgesia. Narcotic therapy may be needed for pain relief.

Treatment of post infectious encephalomyelitis is supportive. However, corticosteroid treatment may be effective\(^{(61)}\).

**Complications:** The Complications of severe encephalitis include; seizures, increased intracranial pressure and respiratory decompensation\(^{(61)}\).

**Prognosis:** Supportive care and rehabilitation are important after the patient recover.
The prognosis depends on the cause and severity of the illness and the patient’s age. If the clinical illness is severe and substantial parenchymal involvement is evident, prognosis is poor\(^\text{(61)}\).

**ACUTE TRANSVERSE MYELITIS (ATM):**

Transverse myelitis is a neurological disorder caused by inflammation across both sides at one level, or segment, of the spinal cord. The term myelitis refers to inflammation of the spinal cord, transverse simply describes the position of the inflammation, that is, across the width of the spinal cord. Attacks of inflammation can damage or destroy myelitis, the fatty insulating substance that covers nerve cell fibers. This damage cause nerves system scars that interrupt communication between the nerves in the spinal cord and the rest of the body\(^\text{(62)}\).

Symptoms of transverse myelitis include Loss of spinal cord function over several hours to several weeks. What usually besines as a sudden onset of lower back pain, muscle weakness, or abnormal sensation in the toes and feet can rapidly progress to more severe symptoms, including paralysis, urinary retention, and loss of bowel control. Although some patients recover from transverse myelitis with minor or no residual problems, others suffer permanent impairments that affect their ability to perform ordinary tasks of daily living. Most patients will have only one episode of transverse myelitis; a small percentage may have a recurrence\(^\text{(62)}\).

The segment of the spinal cord at which the damage occurs determines which parts of the body are affected. Damage at one segment will affect function at that segment and segments below it.

Transverse myelitis occurs in adults and children, in both genders, and in all races. No familial predisposition is apparent.
A peak in incidence rates appears to occur between 10 and 19 years and 30 and 39 years \(^{(63)}\).

The inflammation that causes such extensive damage to nerve fibers of the spinal cord may result from viral infection, abnormal immune reactions, or insufficient blood flow through the blood vessels located in the spinal cord. Transverse myelitis also may occur as a complication of syphilis, measles, lyme disease and some vaccinations, including those for chicken pox and rabies. Cases in which a cause cannot be identified are called Idiopathic \(^{(63)}\).

In post-infections cases of transverse myelitis, immune system appears to play an important role in causing damage to spinal nerves \(^{(63)}\).

Normal MRI has previously been reported in 7-50% of cases \(^{(64)}\).

As with many disorders of the spinal cord, no effective cure currently exists for people with transverse myelitis. Treatment is designed to manage and alleviate symptoms. Therapy generally begins when the patient first experiences symptoms \(^{(65)}\).

**MULTIPLE SCLEROSIS:**

**Background:** Multiple sclerosis (MS) is an inflammatory, demyelinating disease of the central nervous system (C.N.S) \(^{(66)}\). In fact, only 1 of every 4 MS attacks is associated with a viral infection \(^{(67)}\).

The disease can present in different forms, such as primary progressive, relapsing remitting, relapsing progressive, and secondary progressive phenotypes. Genetic susceptibility factors may play a role, as the disease is more common in Caucasian populations living in northern latitudes \(^{(68)}\).

Multiple sclerosis may present in an acute and clinically fulminate form (termed Marburg variant of MS) or may present with concomitant optic nerve
involvement and necrotizing myelopathy (i.e. neuromyelitis optica or Devic's syndrome, considered by some to be an MS variant) \(^{(69)}\).

MS may present in various forms. Some patients have a predominance of cognitive changes, while others present with prominent ataxia, hemiparesis or paraparesis, depression, or visual symptoms \(^{(69)}\).

Optic neuritis presents clinically as orbital pain, at rest or during eye movement, and loss of vision. Patients may complain of patchy loss of vision and upon examination, central scotoma and an afferent pupillary defect may be found. Patients may experience color desaturation even with normal visual acuity, usually manifested as the perception of red color as different shades of orange or gray \(^{(70)}\). Patients with MS may present with facial palsies or trigeminal neuralgia. In fact, the presences of bilateral facial weakness or trigeminal neuralgia strongly suggest the diagnosis of MS. Facial myokymia also may be a presenting symptom. Nystagmus (direction–changing) and internuclear ophthalmoplegia are other manifestations \(^{(70)}\).

**C.S.F:** Oligoclonal bands are distinct electrophoretic patterns that reflect substantial elevation of IgG produced by a restricted set of plasma cells and are demonstrated in CSF samples of approximately 85% of patients with MS \(^{(71)}\).

MRI of head or spine, with and without gadolinium, should be performed according to clinical suspicion for lesion localization.

Typical MS lesions appear as T2 hyperdensities in the ? periventricular region, they have an ovoid appearance, and they involve only the white matter \(^{(72)}\).

Evoked potential testing (visual, auditory, or somatosensory) is especially helpful in detecting clinically silent lesions and documenting an organic basis for range complaints. The most sensitive evoked potentials (50-80% sensitivity) followed by the somatosensory potentials (50-70% sensitivity).
**Medical care:** Thus, preventing disease progression by using available medications is imperative in MS treatment, especially for patients who have been diagnosed early and probably will respond to treatment.

Prevent disease progression by using the “ABC” immunomodulatory drugs (i.e. interferon beta-1a [Aronex], interferon 1b [Betaseron]) \(^{(73)}\).

**Acute exacerbations:** Highly effective treatment is intravenous (IV) methyl prednisolone, Ig for 3-5 days. This medication may help expedite the timing of recovery but will not affect the actual degree of recovery. High dose steroid may work more effectively than oral steroids for the acute attack, and home IV therapy is recommended if the patient does not require hospitalization. Alternatively, high dose oral methyl prednisolone should be used, when feasible \(^{(74)}\).

Azathioprine also may be used as immunosuppressive treatments for MS \(^{(75)}\).

**HYPOKALEMIC PERIODIC PARALYSIS:**

Hypokalemic periodic paralysis causes episodic weakness which usually affect proximal limb muscles more than distal ones; rarely ocular, bulbar, or respiratory muscle are affected. Respiratory muscle weakness may prove fatal. Meals rich in carbohydrate or sodium can provoke attacks. \(^{(76)}\).

Hypokalemic periodic paralysis may complicate thyrotoxicosis in oriental races which corrected by reversal of the hyperthyroidism. \(^{(77)}\).

Reflexes become hypoactive and cardiac arrhythmias may occur during attacks owing to low serum potassium. Men are more often affected because of decreased penetrance in women \(^{(76)}\).
Diagnosis is established by demonstrating a low serum potassium level during a paralytic attack and by excluding secondary causes of Hypokalemia \(^{76}\).

The acute paralysis improves following the administration of potassium salts. Oral potassium chloride (0.2-0.4 mmol/kg) should be given to patients with severe weakness and repeated at 15-30 minutes intervals depending on the response of the ECG, serum potassium and muscle strength \(^{76}\).

**MYASTHENIA GRAVIS (MG):**

*Background:* MG occurs at all ages. Sometimes in association with a thymic tumor or thyrotoxicosis. It is commonest in young women with HLA-DR3; if thymoma is associated, older men are more commonly affected. \(^{78}\).

The incidence of MG is approximately 1:20,000, affect women more than men by a ratio 3:2. Women in the third decade and men in the fifth decade have the peak incidence. Five to seven percent of cases are familiar however; no Mendelian inheritance pattern is demonstrated. \(^{79}\).

Onset is usually insidious, but the disorder is sometimes unmasked by coincidental infection that leads to exacerbation of symptoms. Exacerbation may also occur before the menstrual period and during or shortly after pregnancy. \(^{78}\).

Symptoms are due to available degree of block of neuromuscular transmission caused by autoantibodies binding to acetylcholine receptors; these are found in most patients with the disease and have a primary role in reducing the number of functioning acetylcholine receptors. Additionally, cellular immune activity against the receptors is found \(^{78}\).
**Symptoms & signs:** Patients present with ptosis, diplopia, difficulty in chewing or swallowing, respiratory difficulties, limb weakness or some combination of these problems (78).

Weakness may remain localized to a few muscle groups, especially the ocular muscles or may become generalized. Symptoms often fluctuate in intensity during the day and this diurnal variation is superimposed on a tendency to longer term spontaneous relapses and remissions that may last for weeks.

Nevertheless, the disorder follows a slowly progressive course and may have fatal outcome owing to respiratory complications such as aspiration pneumonia (78).

Clinical examination confirms the weakness and fatigability of affected muscles. In most cases, the extra ocular muscles are involved, and this leads to ocular palsies and ptosis, which are commonly a symptomatic. Papillary responses are normal. The bulbar and limb muscles are often weak, but the pattern of involvement is variable. Sustained activity of affected muscles increases the weakness which improves after a brief rest. Sensation is normal and usually there are no reflex changes (78).

The diagnosis can generally be confirmed by the response to a short-acting anticholinesterase. Edrophonium can be given intravenously in a dose of 10 mg (1ml), 2 mg being given initially and the remaining 8 mg about 30 seconds later if the test dose is well tolerated; in myasthenic patients, there is an obvious improvement in strength of weak muscles lasting for about 5 minutes. Alternatively, 1.5 mg of neostigmine can be given intramuscularly and the response then lasts for about 2 hours: atropine sulfate (0.6 mg) should be available to reverse muscarinic side effects (78).

**Imaging:** Lateral and anteroposterior x-ray of the chest and CT scans to demonstrate a co-existing thymoma, but normal studies do not exclude this possibility (78).
**Laboratory & other studies:** Electrophysiological demonstration of a decrementing muscle response to repetitive 2 or 3 Hz stimulation of motor nerves indicates a disturbance of neuromuscular transmission.

Assay of serum for elevated levels or circulating acetylcholine receptor antibodies is another approach and has a sensitivity of 80 -90 % (78).

**Treatment:** Medication such as amino glycosides, that may exacerbate myasthenia gravis, should be avoided. Anti cholinesterase drugs provide symptomatic benefit without influencing the course of the disease (78).

Neostigmine, pyridostigmine, or both can be used, the dose being determined on an individual basis. The usual dose of neostigmine is 7.5-30 mg (average, 15mg) taken four times daily. Overmedication may temporarily increase weakness, which is then unaffected or enhanced by intravenous edrophonium.

Thymectomy usually leads to symptomatic benefit or remission and should be considered in patients younger than age of 60 years, unless weakness is restricted to the extra ocular muscles. If the disease is of recent onset and only slowly progressive; operation is sometimes delayed for a year or so, in the hope that spontaneous remission will occur.

Treatment with corticosteroid is indicated for patients who have responded poorly to anticholinesterase drugs and have already under gone thymectomy. It is introduced with the patient in the hospital, since weakness may initially be aggravated, once weakness stabilized after 2-3 weeks or any improvement is sustained, further management can be on an out patient basis (78).

Alternate-day treatment is usually well tolerated, but if weakness is enhanced on the non-treatment day it may be necessary for medication to be taken daily. The dose of corticosteroid is determined on an individual basis, but an initial high daily dose (e.g. prednisolone 60-100mg) can be gradually tapered to a relatively low maintenance
level as improvement occur; Treatment with azathioprine may also be effective. The usual dose is 2-3mg/kg orally daily after a lower initial dose.

In patients with major disability in whom conventional treatment is either unhelpful or contra-indicated, plasmapheresis or intravenous immunoglobulin therapy may be beneficial. It may also be useful for stabilizing patients before thymectomy and for managing acute crises (78).

FRIEDREICH’S ATAXIA:

This autosomal recessive disorder, is relatively common with a prevalence of about 1 in 100,000 (80).

This is the most common form of internal ataxia, comprising one – half of all hereditary ataxia (81).

It can occur in a classic form or in association with a genetically determined vitamin E deficiency syndrome (81).

Symptoms and signs: Friedreich's ataxia presents before 25 years of age with progressive staggering gait, frequent falling and titubation. The lower extremities are more severely involved than the upper ones. Dysarthria occasionally is the presenting symptoms, and rarely progressive scoliosis, foot deformity, nystagmus, or cardiomyopathies are initial signs (81).

The neurological examination reveals nystagmus, loss of fast saccadic eye movements, truncal titubation, dysarthria, dysmetria, and ataxia of extremity and truncal movements. Extensor planter responses (with normal tone in trunk and extremities), absent deep tendon reflexes and weakness (greater distally than
proximally) are usually found. Loss of vibration and proprioceptive sensation occurs. The median age of death is 35 years \(^{(81)}\).

Cardiac involvement occurs in 9 percent of patient with Friedreich’s ataxia \(^{(81)}\).

The primary site of pathology is the spinal cord and the peripheral nerves, slight atrophy of the cerebellum and cerebral gyri may occur \(^{(81)}\).

Sclerosis and degeneration are seen predominantly in the spinocerebellar tracts, lateral corticospinal tract, and posterior columns. Degeneration of the glossopharyngeal, vagus, hypoglossal, and deep cerebellar nuclei is described. The cerebral cortex is histologically normal except for loss of Betz cells in the precentral gyri \(^{(81)}\).

**Treatment:** There is no effective treatment. Symptomatic treatment of scoliosis by orthopaedic intervention and cardiac abnormalities by appropriate medication may prolong survival \(^{(80)}\).

**HUNTINGTON DISEASE:**

It is genetic, autosomal dominant, degenerative brain disorders. Frequency 10/100,000. The two clinical hallmarks of the disease are chorea and behavioral disturbance \(^{(82)}\).

The average age at onset is 35 to 45 years. Personality changes, memory loss, and attention deficits are common signs. Depression is also prominent in many patients. The autosomal dominant inheritance is fully penetrante and confers a 50% risk of passage to off spring of affected individuals \(^{(83)}\).

Neuropathologically, the disease predominantly strikes the striatum. Atrophy of caudate nuclei can be visualized in the middle and late stages of the disease \(^{(82)}\).
There is gliosis and neural loss, especially of medium sized neurons in the caudate and putamen \(^{(82)}\). Also there is neural loss in the fourth and fifth layer of the cerebral cortex of the frontal lobe \(^{(84)}\).

Diagnosis of Huntington’s disease has been simplified since a direct genetic test became commercially available. a (CAG) repeat number higher than 39 in the Huntington’s disease gene is diagnostic \(^{(85)}\).

CT-scan usually demonstrates cerebral atrophy of the caudate nucleus in established cases \(^{(86)}\).

If the patient presents solely with progressive intellectual failure, it may not be possible to distinguish Huntington’s disease from other causes of dementia unless there is a characteristic family history or adyskinesia develop \(^{(86)}\).

At present, there is no treatment to prevent the progression of Huntington’s disease. Patients are given symptomatic treatment for the chorea as clorazepam \(^{(85)}\).

CHARCOT-MARIE-TOOTH (CMT) DISEASE:

Several distinct varieties of CMT disease can be recognized. There is usually an autosomal dominant mode of inheritance, but occasional cases occur on sporadic, recessive or X-linked basis \(^{(87)}\).

The responsible gene is commonly located on the short arm of chromosome 17 and less often showed linkage to chromosome 11 or X-chromosome \(^{(87)}\).

Hereditary motor and sensory neuropathy (HMSN) type I and type II, usually present during childhood or adolescence with difficulty in walking or because of foot deformity. The deformity is most commonly pes cavus associated with clawing of the toes and sometimes with equinovarus position of the foot. Muscle weakness tends to
affect the lower leg muscles and may give rise to a bilateral foot drop with a steppage gait. The muscle wasting is often restricted to below the knee producing stork leg appearance \(^{(87)}\).

Weakness and wasting of the small hand muscles appear later. The tendon reflexes become depressed or lost and there is a variable degree of distal sensory loss.

Sensory loss and ataxia tend to be greater and generalized tendon areflexia is usual. Weakness in the hands appears earlier. The peripheral nerves may be thickened. Cases with ataxia are sometimes referred to as the Roussy-Levy syndrome. They are not genetically distinct. The onset in type II form is most often in the second decade but it may be delayed until middle or even late adult life. Some families with type II show genetic linkage with the Duffy blood group \(^{(87)}\).

**SYRINGOMYELIA:-**

Syringomyelia is the development of a fluid filled cavity or syrinx within the spinal cord. Hydromyelia is a dilatation of the central canal by CSF and may be included within the definition of Syringomyelia \(^{(88)}\).

*Types of Syringomyelia include the following:-*

Syringomyelia with fourth ventricle communication is representing about 10% of cases.

Syringomyelia due to spinal cord injury: fewer than 10% of syringomyelia cases are of this type \(^{(88)}\).

*History:* Syringomyelia usually progress slowly, the course may extend over many years \(^{(89)}\).
Sensory, syrinx interrupts the decussating spinothalamic fibers that mediate pain and temperature sensibility, resulting in loss of these sensations, while light touch, vibration, and position senses are preserved. When the cavity enlarges to involve the posterior columns, position and vibration senses in the feet are lost; astereognosis may be noted in the hands. Pain and temperature sensation may be impaired in either or both arms or in a shawl like distribution across the shoulders and upper torso anteriorly and posteriorly. Motor syrinx extension into the anterior horns of the spinal cord damages motor neurons and causes diffuse muscle atrophy that begins in the hands and progresses proximally to include the forearms and shoulder girdles. Claw hand may develop\(^{(89)}\).

An anatomic, impaired bowel and bladder function usually occurs as a late manifestation. Sexual dysfunction may develop. Horner syndrome may appear, reflecting damage to the sympathetic neurons in the intermediolateral cell column\(^{(80)}\).

Asyrinx may extend into the medulla, producing asyringobulbia. This syndrome is characterized by dysphagia, nystaymns, pharyngeal and palatal weakness, asymmetric weakness and atrophy of the tongue, and sensory loss involving primarily pain and temperature sense in the distribution of the trigeminal nerve\(^{(89)}\).

**Physical:** Arm reflexes are diminished early in the clinical course. Lower limb spasticity, which may be asymmetrical, appears with other long – tract sign such as paraparesis, hyper reflexia, and extensor plantar responses. Dissociated sensory impairment may be noted. The syrinx may extend into the brainstem, affecting cranial nerves or cerebellar function. Brainstem signs are common in syringomyelia associated with Chiari malformations\(^{(88)}\).

MRI examination should include sagittal and transverse views in T1 and T2 images proton density sans also can be helpful\(^{(90)}\).

MRA: can be especially helpful in cases of syringomyelia associated with vascular lesions\(^{(90)}\).
Surgical care: Sub-occipital and cervical decompression, laminectomy, syringoитomy, Ventriculoperitoneal shunt, lumboperitoneal shunt and syringoperitoneal shunt (91).

ARNOLD-CHIARI MALFORMATION:

More than half of all cases of syringomyelia are associated with Chiari malformation (92).

Classification:

Type-I: It is characterized by downward displacement of the cerebellum with elongation of the medulla such that the cerebellar tonsils egress through the foramen magnum into the cervical spinal canal. It is not associated with other nervous system malformation (93).

Type-II: By definition; this type is associated with a lumbosacral myelomeningocele and with numerous other nervous system malformations (93).

Type-III: An occipital encephalocele with protrusion of cerebellar remnants into the overlying sac (93).

Type-IV: Isolated hypoplasia of the cerebellum not associated with other nervous system malformations (93).

In patient with Arnold-Chiari malformation, there are commonly skeletal abnormalities on plain x-rays of the skull and cervical spine. CT scans show caudal
displacement of the fourth ventricle. MRI or positive myelography may demonstrate the malformation itself. Focal cord enlargement is found at myelography or by MRI in patients with cavitations related to past injury or intramedullary neoplasm\(^{(93)}\).

MRI scan accurately identify syrinx cavities and associated spinal cord enlargement\(^{(92)}\).

Treatment of Arnold-Chiari malformation with associated syringomyelia is by suboccipital craniotomy and upper cervical laminectomy, with the aim of decompressing the malformation at the foramen magnum\(^{(93)}\).

**Objectives:**

To study the clinical pattern and the aetiology of non traumatic quadriparesis in Sudanese patients.
PATIENTS & METHODS

METHODOLOGY:

- **Study design:** Descriptive cross sectional.
- **Study field:** Alshaab Teaching Hospital.
- **Department of neurology**
- **Study population:** patients with all four limbs weakness. Admitted to the hospital from Jan 2002 - Oct 2003.

**Inclusion criteria:**
1. Adult Sudanese patients with four limbs weakness.

**Exclusion criteria:**
1. Patient below 18 years of age.
2. Patient with traumatic quadriplegia.
3. Psychogenic quadriplegia

**Sampling:**
- **Sample size:** 100 patients.
- **Sample design:** Systematic random sample.
- **Sample frame:** List by registration number.

**Tools of data collection:**
Data were collected by:
1. Pre-design questionnaire.
2. Clinical examination.
3. Investigations.

**Data entry & analysis:**
Data was introduced into the computer from a master sheet recording. Using S.P.S.S. software program, data were entered and analyzed using the student t-test.
- **Age was grouped into:**
  18-24, 25-34, 35-44, 45-54, 55-64, 65-74, >75.

- **Residence was classified as:**
  Khartoum state, central, west, east, north and south.

- **Symptoms considered as:**
  1-Antecedent event in form of:
    Fever, headache, neck pain and backache.
  2-Presenting complains:
    Weakness, sensory complaint, sphincteric dysfunction,
    cranial nerves and cortical symptoms.
  3-Physical signs were grouped into;
    General examinations, systemic and neurological.
  4-Neurological examinations were further arranged into:
    Higher mental function, cranial nerves, upper limbs, trunk and
    lower limbs.
  5-Work up as:
    General and specific investigations.
  6-The specific investigations:
    Imaging, CSF, NCS, EMG and CPK level.
RESULTS

- **Table 1** showed that patients of age group distribution 18-24 to be (21%). 25-34, 13%. 35-44, 21%. 45-54, 13%. 55-64, 16%. 65-74, 13% and > 75, 3%.

- **Table 2** showed the antecedent events, fever was reported in 25%, headache in 14%, neck pain in 16% and backache in 25%.

- **Table 3** showed weakness was reported in all (100%) patients, sensory symptoms were reported by 30% and cranial nerves symptoms were reported by 21%.

- **Table 4** showed that 4% of patients had cortical symptoms, 14% had urinary sphincteric dysfunction and 2% had bowel sphincteric dysfunction.

- **Table 5** showed that cervical cord compression was the final diagnosis in 26% followed by A.I.D.P. 16%, C.I.D.P. 15%, muscular dystrophy 10%, M.N.D. 10%, S.O.L. 4%, dermatomyositis 3%, 2% for each of neurobrucellosis, bilateral infarct, encephalitis and A.T.M. 1% for each of M.S, C.M.T, MG, hypokalaemic periodic paralysis, friedrich's ataxia, Huntington disease, Arnold chiari malformation and syringomyelia.

- **Table 6** showed that 56% of patients had abnormal MRI while 13% had normal MRI.

- **Table 7** showed that the levels of the lesion to be UMNL in 37% of patients diffuse LMNL in 47% and myopathy in 16% of patients.
• **Figure 1** showed that 55% of patients are male and 45% are female.

• **Figure 2** showed that 38% of the patients are resident in Khartoum, 28% in central, 19% in the north, 8% in the west, 6% in the east and only 1% in the south.

• **Figure 3** showed that the onset was gradual in 75% of the patients, rapid in 20% and sudden in just 5%.

• **Figure 4** showed that urinary sphincter disturbance occurred in 14% of patients while the remainder had no sphincteric disturbance.

• **Figure 5** showed that the bowel sphincter disturbance occurred in only 2% of the patients while the remainder had no disturbance.

• **Figure 6** showed that 38% of the patients had upper limbs muscles wasting while no wasting was seen in 62% of the patients.

• **Figure 7** showed that 32% of the patients had lower limb muscle wasting, while no wasting was seen in 68% of the patients.

• **Figure 8** showed the state of tone in the upper limb; 47% had hypotonia, 31% had normal tone and 22% had hypertonia.

• **Figure 9** showed the state of tone in the lower limbs; 51% had hypotonia, 30% had hypertonia and 19% had normal tone.

• **Figure 10** showed the state of the reflexes in the upper limbs; 50% of the patients had absent reflexes, 29% had brisk reflexes and 21% had normal reflexes.

• **Figure 11** showed the state of reflexes in the lower limbs; 51% had absent reflexes, 42% of the patients had brisk reflexes and 7% had normal reflexes.

• **Figure 12** showed that the power in the upper limbs was grade 0 in 3% of the patients, grade I in 6%, grade II in 20%, grade III in 46%, grade IV in 19%, while the power in the lower limbs was grade 0 in 9% of the patient, grade I in 6%, grade II in 33%, grade III in 34% and grade IV in 15%.
• **Figure 13** showed that 40% of the patients had equivocal planter response, 33% had extensor planter response and 27% had flexor planter response.

• **Figure 14**, showed that the sensation of the upper limbs was normal in 79% of the patients, decrease in 20% and absent in 1%, while the sensation in the lower limbs was normal in 76% of the patients, decrease in 20% and absent in 4%.

• **Figure 15**, showed the abdominal reflexes were absent in 59% of the patients and were present in 41%.

• **Figure 16**, showed that 97% of the patients could walk with support and 14% could walk independently and 39% were wheel-chair dependent.
TABLE NO-1:-

Age distribution of patients:

<table>
<thead>
<tr>
<th>Age group</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>18-24</td>
<td>21</td>
</tr>
<tr>
<td>25-34</td>
<td>13</td>
</tr>
<tr>
<td>35-44</td>
<td>21</td>
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<tr>
<td>45-54</td>
<td>13</td>
</tr>
<tr>
<td>55-64</td>
<td>16</td>
</tr>
<tr>
<td>65-74</td>
<td>13</td>
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<tr>
<td>&gt;75</td>
<td>3</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>100</strong></td>
</tr>
</tbody>
</table>
**TABLE NO-2:-**

- **Antecedent events:**

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Fever</th>
<th>Headache</th>
<th>Neck pain</th>
<th>Backache</th>
</tr>
</thead>
<tbody>
<tr>
<td>Percent</td>
<td>25%</td>
<td>14%</td>
<td>16%</td>
<td>25%</td>
</tr>
</tbody>
</table>
**TABLE-NO-3:-**

- The presenting symptoms

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Weakness</th>
<th>Sensory</th>
<th>Cranial nerves</th>
</tr>
</thead>
<tbody>
<tr>
<td>Percent</td>
<td>100%</td>
<td>30%</td>
<td>21%</td>
</tr>
</tbody>
</table>
Concomitant cortical & sphincteric dysfunction

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Cortical</th>
<th>Urinary</th>
<th>Bowel</th>
</tr>
</thead>
<tbody>
<tr>
<td>Percent</td>
<td>4%</td>
<td>14%</td>
<td>2%</td>
</tr>
</tbody>
</table>
**TABLE-NO-5**

The different diagnoses:

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cervical cord compression</td>
<td>26</td>
</tr>
<tr>
<td>A.I.D.P</td>
<td>16</td>
</tr>
<tr>
<td>C.I.D.P</td>
<td>15</td>
</tr>
<tr>
<td>Muscular dystrophy</td>
<td>10</td>
</tr>
<tr>
<td>M.N.D</td>
<td>10</td>
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<td>S.O.L</td>
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</tr>
<tr>
<td>Dermatomyositis</td>
<td>3</td>
</tr>
<tr>
<td>Neurobrucellosis</td>
<td>2</td>
</tr>
<tr>
<td>Bilateral infarct</td>
<td>2</td>
</tr>
<tr>
<td>Encephalitis</td>
<td>2</td>
</tr>
<tr>
<td>A.T.M.</td>
<td>2</td>
</tr>
<tr>
<td>Condition</td>
<td>Count</td>
</tr>
<tr>
<td>-----------------------------------------------</td>
<td>-------</td>
</tr>
<tr>
<td>M.S</td>
<td>1</td>
</tr>
<tr>
<td>Hypokalaemic periodic paralysis</td>
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</tr>
<tr>
<td>Myasthenia gravis (MG)</td>
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<tr>
<td>Friedrich's ataxia</td>
<td>1</td>
</tr>
<tr>
<td>Huntington disease</td>
<td>1</td>
</tr>
<tr>
<td>Charcot Marie Tooth</td>
<td>1</td>
</tr>
<tr>
<td>Syringomyelia</td>
<td>1</td>
</tr>
<tr>
<td>Arnold chiari malformation</td>
<td>1</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>100</strong></td>
</tr>
</tbody>
</table>

**TABLE- NO-6**

- The MRI

<table>
<thead>
<tr>
<th>The result</th>
<th>Normal</th>
<th>Abnormal</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Percent</td>
<td>13%</td>
<td>56%</td>
</tr>
</tbody>
</table>

*TABLE-NO-7*

- The level of the lesion

LIX
<table>
<thead>
<tr>
<th>The level</th>
<th>UMNL</th>
<th>Diffuse LMNL</th>
<th>Myopathy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Percent</td>
<td>37%</td>
<td>47%</td>
<td>16%</td>
</tr>
</tbody>
</table>
DISCUSSION

This is the first study carried out in respect to individual etiologies and clinical presentation of quadriparesis in Sudan.

21% of cases in the study lie in the age group of 18 -24, 13% in the age group of 26-34, 21% in the age group of 35-44, 13% in the age group 65-74 and only3% their age more than 75 years. In conclusion, more than half of the patients (55%) lie in the age group of 18-44, who are the most active sector of the community. (Table1).

Males (55%) were more affected than females (45%). (Figure1).

The majority of patients from Khartoum state and the surroundings (38%), followed by the central states (28%), the north states (19%), the west states (8%), the east states (6%) and only1% for the southern states. This can be explained by the migration toward Khartoum and the war in the south, but no significant geographical etiological factor is suspected .(Figure2).

Family history of neurological disease was reported in 1% of the patients, had Huntington disease.

Weakness was the presenting complaint in all (100%) patients, followed by sensory symptoms in form of tingling or numbness in 30%, but examination of the upper limbs sensation showed decrease in 20% and absent sensation in 1%, while the sensation in the lower limbs was decreased in 20% and absent in 4%. (Table 3 ) and ( figure14).

Urinary sphincteric dysfunction was reported in 14%, while bowel sphincteric dysfunction was reported in 2%. ( Table 4 ).
Backache was reported in 25% and neck pain in 16%. (Table 2).

Only 4% of the patients had cerebral symptoms (convulsion and confusion). (Table 4).

21% of patients had cranial nerve symptoms with same percentage had positive sign on examination. The facial nerve is commonly affected in (11%); 8% bilaterally and 3% unilaterally, followed by the optic nerve 4%, the bulbar cranial nerves (9th, 10th, 11th, 12th) 3%, and 1% for the oculomotor cranial nerve. (Table 3).

The power in the lower limbs was grade 0 in 9%, grade I in 6%, grade II in 33%, grade III in 34% and grade IV in 15%. This is compatible with the degree of disability, which was 14% in patients who experienced no disability, 47% in patients who are able to walk with support, and 39% of patients who are wheelchair bound. (Figure 12 &16).

The state of tone in the upper limbs was flaccid in 47%, normal in 31% and spastic in 22%. While the percentage of brisk reflexes were higher in absent in 50%, and normal in 21%. This is because some patients still had brisk reflexes despite hypotonia or normal tone. Figure 8 &10.

Similarly, the state of tone in the lower limbs was flaccid in 51%, normal in 19% and spastic in 30%. While the percentage of brisk reflexes were higher (42%), absent in 51%, and normal in 7%. Also, this is because some patients still had brisk reflexes despite hypotonia or normal tone. (Figure 9 &11).

Cerebellar ataxia was present in 2% of patients diagnosed as MS and Friedreich’s ataxia.

The planter response was extensor in 33% of patients; most of them had cervical cord compression (26%). (Figure 13).
In this study cervical cord compression constitutes the first major cause of quadriparesis (26%). The age distribution of the patients, 43% more than 60 years, 27% between 45-60 years and 30% between 18-44 years. The gender of the patients is approximately equal. The onset of symptoms found to be gradual in 84% of patients. This reflects that the common cause of cervical cord compression is the cervical spondylosis which is insidious in onset. Neck pain was reported in 53%, while backache in 38% and headache in 8%. This reflects that the majority of patients presented with neck pain and limitation of neck mobility. Associated headache occurred in the posterior region which involve upper cervical spine (C2-C4). Sensory symptoms were reported in 30%, urinary sphincteric disturbance was reported in 20%, while bowel sphincteric disturbance was reported in 8%. Examination of the upper limbs showed wasting in 42%, hypertonia in 42%, brisk reflexes in 50% and decrease sensation in 23% of cases. Abdominal reflexes were absent in 85% of patients. Examination of the lower limbs showed wasting in 34%, hypertonia in 69%, brisk reflexes in 77% and decreased sensation in 19% of cases. Planter response was up going in 69% of patients and equivocal in the reminder 31%. This reflects spasticity with exaggerated reflexes and extensor planter responses is the feature of cervical spondylosis as UMNL. The degree of disability of gait showed; the patients walk with support were about 72% and those who need wheel chair were 24%. MRI detects the diagnosis at all patients (100%).

The most common cause of cervical cord compression in this study is cervical spondylosis which was reported in 21 patients followed by Pott’s disease in 2 patients and one patient with spontaneous extradural haematoma, cervical cord tumor and rheumatoid arthritis complicated atlantoaxial dislocation.

In this study AIDP constitute the second cause of quadriparesis (16%). As in many other studies no age group is exempted (94). Male to female ratio is 2.2:1. Fever
was reported by 44% of patients, this reflect the role of infections as antecedent causative factor. 13% of the patients need ICU admission and ventilatory support. This is compatible with a similar study done at India \(^{(95)}\). Urinary sphincteric disturbance reported in 6% \((1/16)\). This reflect no variation from literature reports and this occur lately in the course of AIDP \(^{(18)}\). Facial weakness had occurred in 56%, \((19\%\) unilaterally and 37% bilaterally), while \((9th, 10th, 11th)\) in 6%. This was compatible with literature reports \(^{(12)}\). Reflexes of the upper limbs were absent in 94%. While all \((100\%)\) patients had absent lower limbs reflexes \(^{(12, 16)}\). CSF protein was raised in 75% of patients which is compatible with other studies that showed 65-85% increase in CSF protein \(^{(14)}\).

CIDP constitutes the third cause of quadriplegic (15%). The study showed that male to female ratio is 3:2. No age group is exempted. Upper limb reflexes were absent in 80% of patients, while lower limbs reflexes were absent in 93% of patients. Only one patient showed cranial nerve abnormality in the form of bilateral facial nerve palsy. 94% of the patients had significant high CSF protein \(^{(14)}\). 67% of the patients had done NCS, which confirm the diagnosis in all of them.

10% of the patients in this study were diagnosed as Muscular Dystrophy. 90% of the patients ages less than 35 years. Which reflect the early onset of the disease \(^{(27)}\). Male to female ratio is 4:1. Examination of the upper limbs showed wasting in 40% of the patients, 60% absent reflexes and all patients had normal sensation. While examination of the lower limbs showed 60% of the patients had absent reflexes and 40% had hypertrophy pseudohypertrophy of the calf muscles. All patients showed marked increase in the level of CPK \(^{(27)}\). 90% diagnosed as Limb Girdle Muscular Dystrophy, while 10% diagnosed as Becker Muscular Dystrophy.
10% of the patients in this study diagnosed as MND. Male to female ratio is 1:1. All patients had no history of fever, neck pain cortical symptoms, and bowel and sphincteric disturbances. 20% of the patients had symptoms due to the cranial nerve involvement. The abnormal cranial nerves include (5th, 7th, 9th, 10th, 11th, 12th), this reflects the affection of MND to the motor nuclei of these cranial nerves. Lower limbs examination showed wasting in 90% of the patients, spastic tone in 30%, brisk reflexes in 40% and normal sensation in all patients (100%). Upper limbs examination showed wasting in 80% of the patients, spastic tone in 30%, brisk reflexes in 80%, up going planter in 50% and normal sensation in all patients (100%). This reflects the involvement of both UMNL and LMNL in MND leading to weakness and wasting of muscles without accompanying sensory changes. 50% of the patients did MRI, all of them were found to be normal. 30% of the patients did EMG and the diagnosis of MND was confirmed in all of them.

4% of the patients had SOL, 25% of them had cortical symptoms. MRI elicits the diagnosis in all patients.

3% of the patients had dermatomyositis. All of them were females, two of them had positive rheumatoid factor (RF) and the other one had positive antinuclear factor (ANA). Muscle biopsy confirm the diagnosis in all three patients.

2% of the patients were diagnosed as neurobrucellosis. The two patients are male and had strong history of contact to animals. The spastic tone, brisk reflexes and up going planter reflexes were noticed in the two patients. The diagnosis was highly suggested by high significant titer.

2% of the patients were diagnosed as bilateral cerebral infarction. The two patients were female of middle age and have spastic tone, brisk reflexes and up going planter reflexes. The risk factor in one patient was hypertension for more than 20 years, while the other one was found to have antiphospholipid syndrome.
2% of the patients had viral encephalitis, in which the diagnosis was suggested by EEG.

2% of the patients had acute transverse myelitis (ATM), one patient had given anti-rabies vaccine two weeks previous to the weakness\(^{63}\). The onset of the disease was rapid.

1% of the patients had hypokalemic periodic paralysis. The patient was a young male; develop sudden quadriparesis without any preceding symptoms. He found to be hypotonic; power of grade 0, normal sensation and absent reflexes. The mental function and cranial nerves were normal. Serum potassium was found to be 1.9 mmol/l. The patient recovered after administration of potassium chloride intravenously.

1% of the patients had MG. the patient was a male, middle age, developed rapid course of weakness with bilateral ptosis, with flaccid tone, absent reflexes and normal sensation. The diagnosis was confirmed by neostigmine test.

1% of the patients had Friedrich’s ataxia. The patient was young male, gradual onset weakness . No family history of similar condition or neurological disease. The patient had marked wasting of all muscle groups of his upper and lower limbs, pes cavus, bilateral foot drop and ataxia.

1% of the patients had MS. the patient was a female, middle age, develop headache, unsteady gait and loss of vision of her right eye. Examination showed; secondary optic atrophy, absent abdominal reflexes and bilateral extensor planter response. MRI suggests the diagnosis.

1% of the patients had Huntington disease. The patient was a female, middle age, developed involuntary abnormal movement in both upper and lower limbs and dementia. The patient had strong family history of similar condition (two brothers and one sister). MRI showed brain atrophy.
1% of the patients had CMT. The patient was young male, had marked wasting, claw hand and bilateral pes cavus.

1% of the patients had syringomyelia. The patient was young female, presented with weakness, paraesthesia. She had dissociated sensory loss. The diagnosis was confirmed by MRI.

1% of the patients had Arnold Chiari malformation. The patient was middle age female, presented with difficulty in speech with nasal tone. She had impaired gag reflex. MRI confirms the diagnosis.
CONCLUSION

• Male to female ratio was 1.22:1, most of the patients were from Khartoum state and central states.
• Most of the patients present with gradual onset.
• Weakness was the presenting complaint in all patients.
• Fever and backache were the presenting complaint in 25% of the patients.
• Wasting of the upper limbs was seen in more than one-third of the patients, while it was less than one- third in the lower limbs.
• The tone was flaccid in approximately half of the patients.
• The sensation was normal in more than two third of the patients.
• The planter was extensor in one-third of the patients.
• Weakness due to UMNL was reported in more than one-third of the patients.
• More than one-third of the patients were wheel chair bound.
• MRI was of great help in picking up brain and cord pathology.
• NCS and EMG were of great help to pick up the diagnosis of diffuse LMNL and myopathy.
• Cervical cord compression, AIDP, CIDP, MND and Muscular Dystrophy are the major etiologies of quadriparenesis in Sudan.
RECOMMENDATIONS

• Al Shaab Teaching Hospital is the tertiary specialized hospital in the country concerned with the diagnosis and management of patients with neurological diseases.
• It received the load of patients from all the states of Sudan. Inspite of this it so deficient in equipments and diagnostic tools.
• Establish many neurological centers throughout the country, connecting to the international center of neurology at Al Shaab Teaching Hospital, inform of training, workshops, research studies and conferences.
• Patients with neurological diseases need special wards, diagnostic material, rehabilitation centers, psychotherapy and medications.
• Establish societies to help these patients with aiding tools and for psychological support.
REFERENCES


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PATTERNS OF CLINICAL PRESENTATION OF 
NON TRAUMATIC QUADRIPLAPARESIS IN 
SUDANESE PATIENTS

Protocol:
Name:----------------------------------- Age:--------- Sex:------
Residence :( Previous – Current)-------------------------------------
Tribe:------- Job:-------- Marital Status:------ Special Habit----------

Symptoms preceding onset:

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Yes</th>
<th>No</th>
<th>Date</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Fever</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Headache</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Vomiting</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Blurring of vision</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Cough</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Diarrhoea</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Neck pain</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Upper limb weakness</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Backache</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Paraesthesia or Numbness</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>- Unsteadiness</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Trauma</td>
<td></td>
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</tbody>
</table>

Symptoms at onset:

Gradual ---------------- Rapid ----------------- Sudden ---------------

Symptoms of cranial nerve affection:

---------------------------------------------------------------------------------------------------
---------------------------------------------------------------------------------------------------
1-Sensory:

<table>
<thead>
<tr>
<th>Loss</th>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
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<td></td>
</tr>
<tr>
<td>Paraesthesia</td>
<td></td>
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</table>
### 2-Motor:

<table>
<thead>
<tr>
<th></th>
<th>Yes</th>
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</thead>
<tbody>
<tr>
<td>Weakness</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unsteadiness</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stiffness</td>
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</table>

### 3-Sphincter disturbance

<table>
<thead>
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<th>Yes</th>
<th>No</th>
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</thead>
<tbody>
<tr>
<td>Urinary</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Faecal</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Impotance</td>
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<td></td>
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</tbody>
</table>

### Cortical:

<table>
<thead>
<tr>
<th></th>
<th>Yes</th>
<th>No</th>
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</thead>
<tbody>
<tr>
<td>Attend conscious</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Seizures</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Others –specify</td>
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</tr>
</tbody>
</table>

### Other Systems:

Co-morbidity:

<table>
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<tr>
<th></th>
<th>Yes</th>
<th>No</th>
<th>Date</th>
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</thead>
<tbody>
<tr>
<td>S/C</td>
<td></td>
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<td></td>
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<tr>
<td>Trauma</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fever</td>
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<td></td>
<td></td>
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<tr>
<td>D.M</td>
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<tr>
<td>Hypertension</td>
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<tr>
<td>T.B</td>
<td></td>
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<td></td>
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<tr>
<td>Syphilis</td>
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<td></td>
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<tr>
<td>Neurological disease</td>
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<tr>
<td>Drugs</td>
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</table>

**F.H:** Consanguinity

<p>| | |</p>
<table>
<thead>
<tr>
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<th></th>
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</thead>
<tbody>
<tr>
<td>S/C</td>
<td></td>
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<tr>
<td>Neurological disease</td>
<td></td>
</tr>
<tr>
<td>Chronic disease</td>
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</table>

### Physical examination:

**General**

1- Fever
2- Anemia
3- Dehydration
4- Tachypnoea
5- BP
6- Pulse

---

LXXVIII
Neck: 

C.V.S: 

Chest: 

Abdomen: 

Back & spine: 

**Nervous system:**

1- **Mental function test:** Glasgow coma scale ( ).
Mood ( ). Intelligence ( ). Speech ( ).
Comment: 

---

2- **Cranial nerves**
I( ), II( ), III( ), IV( ), V( ), VI( ), VII( ), VIII( ), IX( ), X( ), XI( ), XII( ).
Comment: 

---

3- **Upper limbs**
   **Inspection** (Bulk, atrophy, posture, abnormal movements) ---
   **Tone** --------------------------
   **Power** (Grade) ---------------
   **Reflexes:** 2-increased, 1-normal, 0-absent
      Supinator -------------- Biceps -------------- Triceps -------
   **Sensation:** 2-normal, 1-decreased, 0-absent
      Light touch ------------ Pin brick --------------
      Joint position -------------- Vibration -------
   **Co-ordination** ---------------

4- **Trunk**
   **Abdominal reflexes**
      Present  Absent
      Upper quadrant  ------  ------
      Lower quadrant  ------  ------

---
5- Lower limbs

**Inspection** (Bulk, atrophy, posture, abnormal movements) -------
**Tone** ----------------------------------------
**Power** (Grade) -----------------------------
**Reflexes:** 2-increased, 1-normal, 0-absent
  Ankle ------------------------------- Knee -------------------------------
  Planter response ------------------ Clonus -----------------
**Sensation** 2-Normal, 1-Decreased, 0-Absent
  Light touch ---------------- Pin brick ------------------
  Joint position ------------------ Vibration -----------------
**Co-ordination** -------------------------------

6- Others:

**Skull** ----------------------------------------
**Gait** ----------------------------------------
**Type** (Walk independent, with help, cannot walk) -----------------

7- Investigations:

- CBC.
- ESR.
- RBS.
- B. urea & electrolyte.
- VDRL.
- Urine general.
- Stool general.
- Mantoux test.
- CXR.
- CSF.
- Cervical X-ray.
- CT-Scan.
- MRI.
- NCS.
- EMG.