# *ADHESIVE ARACHNOIDITIS*

*Written on behalf of* **COFWA** *by:*

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*Patron: Circle Of Friends With Arachnoiditis*

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**INTRODUCTION:**

Adhesive arachnoiditis is an incurable inflammatory condition affecting the middle (arachnoid) layer of the meninges (which are the membranes surrounding the spinal cord). This condition is thought to be rare, although the real scale of the problem remains yet unknown.

**TERMINOLOGY:**

**Meninges:** 3 membranes which encase the spinal cord and brain. The inner layer= pia mater; middle= arachnoid (has a web-like appearance); outer= dura (tough) mater.

**Arachnoiditis:** inflammation of the arachnoid layer of the meninges. Mild forms often do not cause significant symptoms and may thus go undetected.

**Subarachnoid space:** the potential 'space' between the arachnoid (middle) and pia (inner) meninges; it contains the cerebrospinal fluid (CSF) which flows around the brain and spinal cord, providing it with nutrients and oxygen.

**Epidural fibrosis:** (also: peridural/extradural) scar tissue outside the meninges (literally: outside the dura, the outer layer of the meninges).

**Intrathecal:** this term denotes a site inside the dura; i.e. inside the thecal sac, which is another term used to describe the 3 layers of the meninges.

**Adhesive arachnoiditis:** the most severe type of arachnoiditis, causing scar tissue to form, which compresses nerve roots and impairs their blood supply, leading the various symptoms as a result (see below). Scar tissue may impede the normal flow of CSF.

Note that often in cases of arachnoiditis, scans may reveal epidural fibrosis, and it may well be that the converse is also true, although this is not often acknowledged by medical personnel.

**Important note:** for the purposes of clarity and brevity, this article will refer to adhesive arachnoiditis as AA. This is the clinically significant form of the condition.

**CAUSES:**

The majority of AA cases arise iatrogenically, that is, are caused by medical intervention.
It is helpful to divide the causes into 3 main groups:

1. **Chemically induced AA (CIAA):**
   This arises when chemicals are introduced into or around the subarachnoid space.
   - **Myelogram:** oil-based (Pantopaque/Myodil) and water-based: Metrizamide, Dimer-X, Omnipaque, Amipaque. Procedure used as a diagnostic tool before availability of MRI scans, still in use occasionally. Oil-based dyes remain in the central nervous system as either a thin film or as encapsulated deposits, commonly in the lumbosacral region or in the base of the skull (basal cisterns).
   - **Epidural /intrathecal steroid injection:** therapeutic measure commonly used in both acute and chronic back pain cases, including prolapsed discs. Benefit is questionable and temporary (up to 2-3 months). Risk of arachnoiditis is controversial; evidence of toxicity of the preservatives in the preparation points to a need to reappraise the continued clinical use of this procedure. Preservative-free solutions (Celeston Soluspan/Decadron) may confer lower risk, but this invasive treatment remains one in which risk may well outweigh benefit.

- **Epidural anaesthetics:** again, a controversial subject; use in healthy obstetric patients to minimise pain during labour may be unwise if there are suitable non-invasive alternatives; combined spinal/epidural procedures involve placement of the anaesthetic agent directly into the spinal fluid. Again, it is the preservatives which are likely to cause toxic damage to nerve roots, although the anaesthetic agents themselves may also directly affect nerves. The practice of regional anaesthetic techniques such as epidurals in conjunction with a general anaesthetic (used in paediatric operations) is a cause for considerable concern as the patient is unconscious and cannot therefore alert the doctor performing the procedure to pain due to inadvertent injection directly into nerve roots. Note also that in procedures of epidural steroid injections, it is common practice to combine this with local anaesthetic to confer immediate relief (steroid aiming to provide a more sustained relief over weeks): thus conferring "double jeopardy".
- **Chymopapain:** this agent has been used as a chemonucleolytic agent; it is an enzyme, which breaks down disc material that has leaked due to a disc herniation (prolapse).
- **Intraspinal chemotherapy agents:** e.g. methotrexate which is used to treat certain cancer conditions and is deemed to provide higher available drug concentrations than if given intravenously; however some authors have suggested that it is unnecessary to use intraspinal injections.
- **Chemical meningitis:** may result from any of the above procedures; it involves acute inflammation of the meninges, often in both the spinal and cerebral (around the brain) areas.

2. **Mechanically-induced AA (MIAA):**

- **Spinal surgery:** especially multiple surgeries.
- **Trauma**
- **Multiple lumbar punctures**
- **Spinal stenosis** (when chronic)
- **Anatomical abnormalities:** especially degenerative conditions: e.g osteophytes (bony protuberances)
- **Chronic disc prolapse:** including leaked disc material, which is known to be highly irritant to nerves.
- **Blood:** bleeding into the spinal fluid due to invasive procedures or trauma (as above). Blood is extremely irritant to nerves. Subarachnoid haemorrhage may occur spontaneously (no invasive procedure precedes it) and can cause arachnoiditis.

3. **Infection:**

- **Meningitis:** viral/bacterial; inflammation in the meninges; usually cerebral, but may also affect the spinal meninges. Lumbar puncture is required to establish a diagnosis.
- **Tuberculosis:** before the advent of widespread myelograms etc. TB was a major cause of spinal AA, often in the thoracic (chest) region of the spine. There has been some increase in the incidence of TB in Western countries in recent years, possibly due to immigration from India and Pakistan, where TB is still common. TB of the spine (Pott's disease) remains relatively uncommon though.

**Note:** observations of an anecdotal nature, arising from over 3 years of regular communication with AA sufferers around the world have led to the following observations:

1. CIAA tends to cause a more florid, severe condition with widespread symptoms; in the cases where multiple chemical insults have been sustained, there is a more severe picture, so that one can postulate that the severity of the condition is proportional to the number of invasive chemical procedures have been undergone.
2. MIAA tends to cause more localised damage, which affects one, or two nerve roots and causes symptoms related to this specific damage.
3. Commonly, patients with AA will have undergone a variety of medical procedures, the condition being multifactorial in origin. This gives rise to problems with regard to attempted litigation. Further

investigation comparing CIAA and MIAA needs to be undertaken in order to discern a workable clinical picture, which may be useful both in diagnostic terms and within a legal framework.

**SYMPTOMS:**

AA does not have a typical clinical presentation, although there are a number of features, which are common in people with the condition. However, the picture is somewhat complicated by the fact that the symptoms of AA occur against a backdrop of the original spinal problem for which invasive procedures were undertaken (except in a small minority in which no spinal condition has occurred, for instance, in AA secondary to epidural anaesthesia in childbirth).

It is important also to remember that a number of the symptoms experienced are common to various chronic illnesses and may well arise secondary to the general debility occasioned by unrelieved pain and stress resulting from dealing with illness that is relentless for years on end.

Chronic pain is not regarded by most of the medical profession as detrimental of itself; however, recently some doctors are beginning to voice a different point of view, recognising that unrelieved pain constitutes a source of constant stress on the body, resulting in over-production of stress response chemicals in the body, such as adrenaline, insulin and cortisol. These substances cause a variety of problems. In America, highly sophisticated PET scans have shown that chronic pain in some way alters the way in which the brain responds to stress or pain; the concentration of neurotransmitters (chemical nerve messengers) in certain brain areas seems to vary from that of healthy people.

In 1999, a global postal survey of people with arachnoiditis showed the following results:

1. Pain (100%)
2. Numbness/tingling (86%)
3. Sleep disturbance (84%)
4. Weakness (82%)
5. Muscle cramps/twitches/spasms (81%)
6. Stiffness (79%)
7. Fatigue (76%)
8. Joint pains (72%)
9. Balance difficulties (70%)
10. Loss of mobility (68%)

Other common symptoms seen in the typical case:

1. Bladder/bowel/sexual dysfunction(68%)
2. Increased sweating (63%);
3. Difficulty thinking clearly/Depression (63% /62%);
4. Heat intolerance(58%);
5. Dry eyes/mouth(58%) and
6. Weight gain (50%).

Heartburn/indigestion is also a common problem; often this is related to use of NSAIDs (anti-inflammatory drugs). Difficulty in swallowing may be related to this or may arise (less commonly) due to inco-ordination of the gullet muscles.

Headaches are also a common feature. Many people seem to develop skin rashes, for unclear reasons. (some may be related to medication such as anticonvulsants).
Other less common problems experienced include: Tinnitus (ringing in the ears), dental problems (tooth decay may be worsened by dry mouth due to loss of the protective power of saliva), abnormalities in the menstrual cycle, eyesight problems (difficulty in focussing may be due to medication).

The pain tends to be intractable and resistant to treatment, being predominantly neurogenic in origin. This causes persistent burning pain and intermittent stabbing or electric shock type pains. Burning in the feet is common and may be accompanied by a sensation of walking on broken glass.

There may also be a component of central pain, which is well known to be difficult to treat. This involves various bizarre sensations, such as pain felt on light touch or a change in temperature (allodynia) or pain felt in a different part of the body to the one being touched. People also experience sensations such as water running down the leg, or insect bites.

One doctor has likened the pain of AA to that experienced in cancer, but without the relief of death. Indeed, some sufferers become suicidal due to the unrelenting pain and the neurological deficits they experience.

There are a range of systemic symptoms which constitute a debilitating condition that severely impairs the sufferers' quality of life.
AA is incurable and may be progressive in some cases. Usually people tend to 'plateau out' at a certain level of pain/loss of function, but in a minority, a relatively trivial event such as a slight fall or car accident, can set off a rapid decline.

**Note:** in the survey, a number of respondents had a diagnosis of an autoimmune disorder such as Lupus, Sjogren's, Rheumatoid arthritis. There appears to be a possible link between AA and autoimmune type problems. Out of 317 survey respondents, 27 had thyroid disorders, all except one having previously undergone myelography. As myelogram dyes contain iodine, there may be a significant link between the myelogram and subsequent thyroid disease; this is currently being investigated. There are also a number of arachnoiditis patients who have also been diagnosed with Multiple Sclerosis, as well as several more who have undergone investigation for MS. Those who have a diagnosis of fibromyalgia in addition to arachnoiditis are probably suffering from the condition as a secondary feature of the underlying arachnoiditis; fibromyalgic type symptoms of diffuse muscle tenderness and fatigue are common in arachnoiditis patients.

**Important note:** not ALL symptoms can be ascribed to arachnoiditis. Any new or increasingly severe symptom which persists for more than 48 hours should be fully assessed at a medical consultation.

**DIAGNOSIS:**

Many people who have symptoms such as those described and a history of risk factors for AA still have difficulty in getting a diagnosis. As the condition is perceived to be rare, doctors often do not consider it a likely diagnosis. It is important that treatable conditions such as recurrent disc herniation are identified and treated and this can be achieved through the use of an MRI scan. High resolution scans may also be able to demonstrate AA, although in the early stages it might not be picked up. In any case, one must bear in mind that MRI scan results often fail to correspond accurately to the clinical picture. Heavy reliance on the need for a diagnosis is unadvisable, and often unnecessary, as management of symptoms is the only option, AA being incurable.

EMG (electromyogram) or NCV (nerve conduction velocity) tests may be performed to assess nerve damage. If there is loss of bladder control, urodynamic studies may be undertaken to fully assess the problem.

**DIFFERENTIAL DIAGNOSIS:**

This refers to other similar diagnoses which may be relevant:

- **Failed Back Surgery Syndrome:** in fact, arachnoiditis probably accounts for over 10% of FBSS cases; FBSS is common, incidence varying from 25% to 40% of all spinal surgery cases. The commonest causes include: epidural fibrosis, recurrent disc herniation, spinal stenosis (narrowing of the spinal canal or the foramina (holes in the vertebrae) through which the nerve roots exit from the spinal cord. It is important that treatable causes such as reherniation of a disc, are identified and treated.
- **Multiple Sclerosis:** as mentioned above, it is quite common for arachnoiditis patients to be investigated for MS.

- **CRPS:** previously termed RSD: reflex sympathetic dystrophy, CRPS Type I refers to problems in one limb, often after trauma/surgery: pain, swelling and changes in skin colour and temperature, abnormal sweating: increased/decreased (bone density lost in later stages). CRPS Type II (previously causalgia) refers to more widespread problems, other than in the area affected by an injured nerve and resembles arachnoiditis. Continuous pain, allodynia (pain from non-painful stimulus such as light touch/clothing/temperature change) and/or hyperalgesia (heightened pain) can occur. (also: skin rashes, abnormal body temperature, tremors (shakes), tripping/falling.)
- **Cauda Equina Syndrome:** acute CES is a surgical emergency; loss of bladder/bowel function, saddle anesthesia (loss of sensation or tingling in the buttocks and around the anus/vagina/genitals), leg weakness and severe pain in the lower back/limbs/genitals. CES is basically a descriptive term for a set of symptoms. It may arise when there is a severe compression in the cauda equina, (horse's tail) at the lower end of the spinal cord (acute causes include large disc prolapse). A chronic equivalent to CES may arise in arachnoiditis.

**MANAGEMENT:**

As explained above, AA is incurable, but there are a number of measures which may be helpful in managing symptoms.
Sadly, in a survey in 2000, I found that quite a high proportion of AA patients continue to experience very troublesome levels of pain as well as other symptoms including loss of function.

In part, this may be due to reluctance of medical personnel to prescribe medication in the long term, especially narcotic painkillers, which are perceived as carrying a high risk of addiction. In fact, narcotics used for pain relief (in comparison with recreational use) carry a very low risk of addiction in the generally accepted sense of the term. Whilst the body becomes accustomed to a certain level of medication and goes into withdrawal if the drug is abruptly discontinued, psychological dependence is uncommon. Behavior that may be regarded by medical and paramedical practitioners as addictive, may arise out of a desperate bid to find adequate pain relief. Often, doctors are willing to prescribe medication such as anxiolytics or hypnotics (treating anxiety or insomnia) such as Valium (diazepam) or related drugs: which are in fact, far more addictive than narcotics (morphine and related drugs) and carry a risk of tolerance (needing increasing doses) that far exceeds that of narcotics. Usually, what the patient really requires is an increased dose of painkiller to relieve their 'anxiety' or sleep disturbance. Families may also be wary of narcotic use, deeming it as inappropriate; the stigma is still very much in evidence.

Management of AA should revolve around a wholistic approach and may require a multidisciplinary team involvement. However, this should be overseen by one individual amongst the medical personnel (usually the primary doctor or GP) It is vital that the patient develops a working therapeutic alliance with his/her doctor(s). This will pave the way to a good level of compliance and a mutual trust and respect.

Treatments should be implemented one at a time and must be trialled for at least 4-6 weeks (unless there are severe side effects or allergic response) in order for adequate assessment of their efficacy can be made.

Round- the- clock dosing is essential to achieve effective pain relief and minimize side effects and tolerance (need for increasing doses to achieve the same effect).
Usually side-effects begin to subside after about 10-14 days of continued usage, so patients should be advised to ride out the first few days of sedation, nausea etc. if possible. Persistent side effects such as constipation and dry mouth are common, but may be managed fairly easily.

Below is a brief outline of the various strategies which comprise a multimodal programme:

1. **Medication**: often oral, but may also be via a patch. Typically, a triad of narcotic/antidepressant/anticonvulsant is used, +/- muscle relaxant +/- anti-inflammatory. (see below for more detailed list)
2. **Physical therapies :** massage (Shiatsu), chiropractic, cranialsacral therapy, Myofascial Release techniques; stimulating: Low Level Laser Therapy, Ultrasound, TENS; Acupuncture;

3. **Exercise:** loss of mobility may have a knock-on effect in general debility, and can directly contribute to development of osteoporosis; gentle exercise is helpful; 'No pain, No Gain' does NOT apply and exercise regime needs to be carefully tailored to the needs of the individual. Feldenkrais, hydrotherapy, isometric exercises are often helpful.
4. **Treatment of specific problems:** e.g bladder dysfunction; poor circulation in extremities
5. **Management of side effects:** such as constipation
6. **Nutritional:** avoidance of caffeine and possibly trigger foods; supplements such as vitamins, MSM, glucosamine.
7. **Herbal/homoeopathic:** NB. Herbal preparations may interact with prescription medication; St. John's Wort (depression); Gingko
8. **Lifestyle measures:** smoking: preferably should be stopped as it worsens circulation; alcohol: may interact with medication; if taken in excess, as a strategy to aid sleep/reduce pain or distress, it may act as a depressant i.e. causing a low mood. Illicit drugs such as cannabis have been reported as helpful in reducing muscle spasms and enhancing pain relief; cannabis is currently being trialled in the UK for use in MS patients.
9. **Psychological:** often people with arachnoiditis are reluctant to admit to emotional distress because they have been labeled as having a psychosomatic illness in the past; however, psychological difficulties are only to be expected in a debilitating, incurable illness. Often complex psychological situations may arise, particularly with regard to the causative factors (being mostly iatrogenic): anger and bitterness can be very strong and persistent, often being fuelled by day-to-day frustrations over loss of function, relationship troubles (as with any chronic illness, considerable strain is put upon partners and family) and fear for the future. Individual counseling, couples, or group therapy may help address issues on grief (over loss of health, role, financial security, self-esteem etc.etc.) In addition, patients can be instructed on strategies for self-help in learning to cope with ongoing illness and pain; these include Cognitive Behavior Therapy, which can be very helpful
10. **Support groups:** groups are invaluable in allowing sufferers to be in direct contact with others who are going through the same sort of troubles. This contact helps to reduce the strong sense of isolation which is extremely prevalent in people with chronic illness.
11. **Information:** the Internet can be a very useful resource, but it must be remembered that not all the information is from reliable sources; one should always check that material uses reputable (and verifiable) references. Support groups can be sources of useful information on the condition and other issues regarding the day-to-day effects of the illness on various aspects of life.
12. **Aids:** ranging from simple measures such as pads to place in shoes to make walking more comfortable to wheelchairs; these can really help in daily life.

**Medication:**

The majority of sufferers need to use a variety of medication in an attempt to reduce the pain.

The survey results showed the following as regards treatment regimes and in most cases, polypharmacy( a cocktail of drugs) is necessary:

- OPIATES (e.g. Morphine, Pethidine (Demerol), Methadone, Tramadol etc.): 171=54% (note: Buprenorphine: Temgesic is a partial opiate agonist: and partial antagonist; this means that it may give rise to withdrawal symptoms in patients who have previously taken strong opiate drugs)

- ANTI-INFLAMMATORY (e.g. Brufen, Mobic, Naproxen, Vioxx etc.): 144=45%

- ANTIDEPRESSANT (commonest amitriptyline; also Prozac etc): 90=28%

- ANTICONVULSANT (e.g. Tegretol; Neurontin; Vigabatrin): 84=26%

- MUSCLE RELAXANT: (e.g. Baclofen; Robaxin; Dantrolene; Zanaflex): 34=11%

- BENZODIAZEPINE (e.g. Diazepam, Clonazepam, Nitrazepam, etc.): 39=12%
- DIURETICS (for fluid retention): 17=5%
- INA (intraspinal narcotic agents= "the pump"): 8=2% incl. CLONIDINE: 2
- SCS (spinal cord stimulator): 2
- STEROIDS: 4 (1 via portal implant)
- QUININE (for muscle cramps): 3
- OXYBUTININ (for bladder muscle instability): 1
- BETHANECOL (for urinary retention): 1
- ETIDRONATE (for prevention of bone loss in osteoporosis): 1
- NONE: 10=3% PARACETAMOL/ASPIRIN ONLY: 4
- TENS: 2

Note low percentage on no medication or simple analgesia; generally, for respondents who were not on medication, this was due to inability to tolerate stronger medication due to side-effects or adverse reactions.

Most cases in the survey involved polypharmacy, with a combination of opiates with antidepressant and/or anticonvulsant being common. Anti-inflammatory medication (NSAIDs) usage was common despite a considerable number of respondents stating that they had had to discontinue use due to adverse gastric effects (e.g. gastric/duodenal ulcer, heartburn, gastric bleed), which are well known with this type of medication.

Antidepressant medication is used at a sub-therapeutic dose as regards treating depression (i.e. say 25mg amitriptyline rather than 75-150mg) it is useful for neurogenic pain. Tricyclic antidepressants are most effective, whereas SSRIs (newer type) such as Prozac are often poorly effective. Of course, in some cases, full antidepressant dose may be given to combat any depressive features compounding the physical problems. Anticonvulsant medication is useful for neurogenic pain.

Benzodiazepines: a group of drugs including valium: used either as a muscle relaxant or to combat anxiety, or perhaps as sleeping tablets.
Naturally, high doses of these drugs may cause significant adverse effects such as sedation, cognitive impairment, nausea and vomiting, fluid retention etc.

**PROGNOSIS:**

The outlook for patients with arachnoiditis is unfortunately as yet unknown. There has only really been one medical article written about this, published in the late 80s by Guyer. He contends that on average, life expectancy may be shortened by as much as 12 years. Arachnoiditis, as discussed above, is as yet incurable.

There is considerable controversy over whether it is a progressive condition. In the majority of patients, there may be a gradual decline over a period of years with increasing pain levels and some loss of function. Some people seem to reach a plateau and remain stable. Those who can stay off strong medication and are able to maintain a reasonably active lifestyle seem to do best. In a small minority, a quite minor injury from a fall or car accident can trigger rapid decline.

A few patients go on to develop complications such as arachnoid cysts (encapsulated fluid collections around the spine or in the brain), syringomyelia (fluid-filled cavity in the spinal cord) or hydrocephalus (enlarged

ventricles in the brain). Other complicating conditions include: depression, osteoporosis (due to lack of mobility).
By and large, though, there are AA patients who have had the condition for up to 20 years and are still able to be relatively independent and reasonably mobile.
AA is not directly fatal. However, there have been cases of suicide due to the despair of unrelieved pain.

**THE FUTURE:**

One of the most tragic losses experienced by arachnoiditis patients is hope for the future. Most see a bleak and pain-filled existence centred around an unrelenting illness. However, we must never lose sight of hope: as Bernie Siegel wrote:

" Hope isn't statistical, and individuals recover. There will always be a first person to recover from every disease…..there is no false hope. False hope tends to be a recital of statistics, and people are not statistics. But there is false no hope."

What better note to end on than to quote Dr. Goodling, from Duke University, who said:

**"It's so important for people who are hurting to know that the story hasn't been finished. Things are terrible now, but there's more to the story."**