

## MYALGIC ENCEPHALOMYELITIS (ME) IN CHILDHOOD

### BACKGROUND

The term Myalgic Encephalomyelitis was first used in this country in connection with the epidemic at the Royal Free Hospital in London in 1955. (1) Two hundred cases were documented and the disease was accepted as organic in nature by the physicians involved. The epidemic consisted of a viral meningo-encephalitis which left many of the affected with a more chronic illness. Subsequently and retrospectively (in 1973) two psychiatrists wrote a paper in which they suggested the problem had mainly been due to an outbreak of mass hysteria among nurses and female doctors, the RFH at that time in history having a high preponderance of female doctors and medical students. The authors putting forward this theory did not actually see any of the patients but simply reviewed the case records. (2)

This paper was the start of the process whereby the medical profession retreated into a position of disbelief regarding ME, and allowed the area to be taken over by psychiatry for c 30 years. Fortunately things are finally improving, especially with the publication in 2002 of the CMO's report which concluded that "ME is a genuine illness and patients should not be dismissed as malingerers"

Looking back it turns out that there had been earlier epidemics of an essentially similar illness which occurred in Los Angeles (1934) and Iceland (1947). At present in this country the majority of cases seem to be sporadic.

### DIAGNOSTIC LABELLING

This condition has been called the Disease of a Thousand Names. Some of these are:- Iceland Disease, Tapanui Flu (New Zealand), Neurasthenia, Post Viral Fatigue Syndrome, Chronic Fatigue Syndrome, Chronic Fatigue and Immune Deficiency Syndrome (USA), Atypical Migraine and Post-Polio Syndrome.

### ME IN CHILDHOOD

Over recent years an increasing number of cases have presented in childhood. I have seen approximately 200 children with this condition in Durham over the last 20 years, and a further c300 from outside the area. The condition appears to be increasing in frequency, and is essentially similar to that seen in adults. Most of my cases are teenagers but I have a handful of quite severe cases in the 5-10 age group. I have seen two cases where the onset appeared to be before the age of 2yrs.

### AETIOLOGY

This remains an area of uncertainty. My personal conviction is that ME is **primarily** an organic illness, and that the role of psychological factors in the causation of this illness is around 0%. Obviously like any unpleasant chronic illness there can be **secondary** psychological consequences, but I personally believe that the importance of psychological factors in prolonging the illness has been greatly exaggerated. Unfortunately, in this and many other

countries the medical profession has allowed the condition to be dominated by the psychiatric school of thought to a degree that many sufferers have understandably found quite upsetting. Organic theories of aetiology in this country are heavily weighted towards a post-viral hypothesis, although some workers see the ME syndrome as a final common pathway for multiple disease agents (including toxins/pesticides especially organophosphates). American workers favour an immunological hypotheses, postulating chronic immune activation. More recently the possibility that some cases of ME are caused by Tick-borne diseases (eg Borrelia as in Lyme Disease) has been raised and leads to the possibility of treatment with long-term courses of antibiotics.

The disease is best thought of as a multisystem disorder with an encephalopathic element as this is the only way the wide array of symptoms can be adequately explained.

Work by Costa in London has demonstrated consistent abnormalities of brain stem and cortical perfusion by PET scanning in adult ME patients.

## **DIAGNOSIS**

In a proportion of cases seen early, infection with an enterovirus or influenza can be documented. Some cases follow mononucleosis or hepatitis A. However, in most cases by the time the condition is suspected there are no abnormalities on routine investigation [PET scans not being widely available at present]. Accordingly, diagnosis is essentially CLINICAL, and is based almost entirely on the HISTORY (there being no hard physical signs or specific laboratory tests. One possible exception to this statement is that many sufferers have a characteristic marked facial pallor with a translucent character).

## **CLINICAL PATTERN AND COURSE**

Around 50% of cases start acutely with a severe/infectious process. The remaining cases come on gradually and it may be difficult to date the onset of the disease. Most cases follow a prolonged course with fluctuation in the severity of symptoms and mini-remissions followed by relapses following further infections or stress.

## **PROGNOSIS**

Mild cases tend to recover sooner and more completely. (However, mild cases can be turned into severe cases e.g. by a further nasty infection).

Moderate cases tend to drag on for up to 5 years.

Severe cases can persist into adulthood with a very guarded long-term prognosis. Fortunately however, some severe cases can make complete recoveries.

Sadly, the reality is that at the extreme end of the spectrum of severity there are a small number of sad cases where the young person appears to have actually died from their ME.

## **SYMPTOM COMPLEX**

It is vital that a proper history is taken and that sufficient time is allocated for this task. Only then will the full range of symptoms be uncovered. [The commonest reason for missing the

diagnosis is to concentrate on one symptom only, or to take an incomplete history]. The classical symptoms include the following:-

(i) FATIGUE/FATIGUEABILITY

This is a cardinal symptom and can have different facets. The fatigue is both **Physical** and **Mental**. Mild cases may be capable of significant acts of exertion, but typically will be exhausted for days or even weeks following this. More severe cases will not be capable of the exertion in the first place. When physically tired, patients will notice their mental faculties deteriorate, and they can become confused and forgetful.

**All the other symptoms can be made worse by physical exertion, and mental effort can make the physical fatigue worse.**

[The best description a patient gave was "Its like running a marathon with a bad dose of flu"].

(ii) HEADACHE

This can be severe, continuous, generalised and throbbing and can last for months or years. This is one of the commonest symptoms, occurring in up to 90% of cases.

(iii) ABDOMINAL PAIN  $\pm$  NAUSEA

(iv) LIMB PAINS ("myalgia")

(v) SLEEP DISTURBANCE

This usually consists of hypersomnolence in the acute stage, followed later by sleep reversal, whereby the child finds it difficult to get off to sleep, but then can't wake up the next morning.

(vi) NEUROLOGICAL SYMPTOMS

- a) Problems with memory, concentration, cognition, nominal aphasia
- b) Vertigo
- c) Hyperacusis/photophobia, ie undue sensitivity to noise and light
- d) Paraesthesiae, muscle twitching
- e) Sudden severe attacks of generalised pain ("sensory storms")

(vii) AUTONOMIC DYSFUNCTION

Problems with temperature control, feeling alternately too hot or too cold. Postural hypotension (ie dizziness when standing up).

## MANAGEMENT

Other organic illnesses should be excluded by appropriate investigations (coeliac disease, Addison's disease, thyroid disease, juvenile chronic polyarthritis, Lyme disease.....etc).

While current orthodoxy promotes the use of Graded Exercise regimes (GET) and Cognitive Behavioural Therapy (CBT), my opinion is that these therapies are of only marginal benefit in some mild and convalescent cases and have very little to offer the moderate and severe cases. Indeed, I have seen several cases where over-robust regimes of GET have almost certainly been responsible for marked deterioration in the condition. NICE Guidelines state that there is no evidence that CBT or GET are effective in severe cases.

Accordingly, it should be emphasised very clearly that **currently there is no curative treatment available for ME/CFS**. Patients quite accept this and it is often the doctors who find it difficult, and thereby fall into the trap of the "Therapeutic Fallacy" whereby they can't accept the failure of their treatment regimes; this can lead to them rejecting their patients with ME/CFS or even worse, blaming the patient or family for the failure of their treatment. **I have been involved in >50 cases where children have been subject to Child Protection procedures and threatened with removal from their families because of these (in my view) mistaken belief systems. These cases could be seen as "Child Abuse by professionals").(3)**

I personally start from a position of therapeutic nihilism and take it from there! Then one can accept that, as in any chronic illness, patients deserve lots of support. This comes in the form of the following:

### A) SYMPATHY

Simply making an official diagnosis is very important. In doing so the doctor can show that he/she believes in the genuineness of the condition and this **validation** is the most important step in beginning to help and support the family. Explaining the implications of the diagnosis is not exactly "good news", but it is best to be honest and the potential for bad news should not be glossed-over or minimised. The family need to be warned of the dangers of overdoing things in the early stages which can possibly make things worse. Explain the guarded prognosis and the lack of curative treatment. Then offer sympathy and

### B) SUPPORT

Children with this condition deserve just as much support and help as any child with a chronic illness. In my view, all but the very mildest cases deserve long-term support from a consultant paediatrician, rather than simply being discharged to the care of the G.P.

Practical help can include writing to the school explaining the condition, arranging Home Tuition for the cases too unwell to attend school, getting extra time allowed for exams, and rest periods during exams, helping with DLA claims and appeals and putting in touch with ME support organisations. Young sufferers attending university can be helped by explanatory letters, and support eg for granting residence on ground floors and lodgings close to lecture halls.

### **C) PROTECTION**

Children without a proper diagnosis of ME are at risk of being wrongly diagnosed as malingering or school phobic by their parents, teachers, peers and assorted professionals. The most important step in protecting them from this nightmare scenario is that they be officially diagnosed as ME. It is important to get all members of the family "onside" as it can be very upsetting for the child if eg a distant granny or non-custodial parent adopts a policy of disbelief.

In the challenging area of the very severe cases, I have found the young person needs protection from too many well-meaning professionals as they can actually add to the stress rather than helping. Often the paediatrician panics and refers to psychiatry, who then label the child (wrongly), not as severe ME but as having the "Pervasive Refusal Syndrome", and subject the child to inappropriate regimes.

It is this area that in my opinion justifies the use of the term **ME** rather than CFS because of its power and strong aura of organic illness. Children and families need this power to help them cope with the above threats. The term "Chronic Fatigue Syndrome" is mealy-mouthed by comparison, and cuts no ice in the playground when a child returns to school after a gap of nine months!

(In the memorable words of one campaigner, Tanya Harrison of BRAME, in her evidence to the CMO's working Party, "the label Chronic Fatigue Syndrome is as unhelpful to ME sufferers as the term "Chronic Forgetfulness Syndrome" would be to families and patients with Alzheimers' Disease"!)

### **D) SYMPTOMATIC TREATMENT**

There is quite a bit doctors can do to help with symptoms such as pain, sleep problems, etc. I have found drugs like amitriptyline helpful for sleep reversal and pain, and Melatonin can also be very effective for sleep although its beneficial effects seem to wear off after a few months. More recently I have found the use of "Ritalin" (methylphenidate, a drug used in ADHD) quite gratifyingly effective in treating the "brain fog" of ME, often making a crucial difference to the ability to benefit from home tuition and to perform and survive in examinations.

Many severe cases suffer from poor appetite and nausea and in a small number of cases one has to resort to tube feeding for a period of time. They are at risk of being labelled as cases of Eating Disorder and again referred to psychiatrists.

Naturally if psychological symptoms/ depression are prominent then these should be managed on their merits. If referral to Psychiatry is being offered, make it clear that this isn't in any way an act of disbelief in the genuineness of the condition. Similarly, avoid referral to "disbelieving" psychiatrists in the first place as such consultations can be very upsetting for the young person and family.

Naturally, the involvement of families with ME organisations both local and national is exceedingly important. I have found in my area the support given by our local group absolutely invaluable and it has made my job of supporting the families that much easier. (This is another

justification for using the term ME, as all the support organisations use it, and there isn't a charity for Chronic Fatigue Syndrome"!) )

Good Luck to all sufferers, their families and the professionals caring for them!

### **Stop press 2011**

A study from Bergen, Norway (3) has found dramatic beneficial effects from the use of Rituximab, a monoclonal antibody to B cells. In a randomised control trial, 10 out of 15 patients treated experienced major improvements. Further trials are shortly to be conducted.

2017 Unfortunately these further trials proved negative, although there may still be a subsection of patients who would respond

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