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THE LATE EFFECTS OF ME - Can they be distinguished from the Post-Polio Syndrome?

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Introduction:

Few people would dispute that ME (Myalgic encephalomyelitis), an illness which blights the hopes and aspirations of all sufferers, especially the young, is denied equal treatment in respect of diagnostic facilities, medical coverage and welfare provision. Comparable chronic and unpredictably disabling neurological conditions, for example Multiple Sclerosis, which was formerly ascribed to "hysteria" and similarly neglected, now receive government recognition, facilities within the NHS, and more generous research funding - though the potential cost of effective treatment can still arouse bitter debate.

What is ME?[1,2,3]

a. ONSET:

It is a syndrome (a group of linked symptoms) initiated by one or more of a related group of enteroviruses which circulate annually in the community in summer and autumn in temperate climates, but all the year round in tropical areas.

b. MINOR ILLNESS:

The majority of encounters with these viruses are asymptomatic but some subjects, more commonly teenagers and adults, suffer a seemingly trivial minor illness, usually described as a non specific summer 'flu accompanied by gastrointestinal upset, sore throat and occasionally by generalised glandular enlargement.

c. SECONDARY PHASE:

The minor illness is self limiting in 90% of adults. However, some 5-10% of all age groups exposed, may progress to a more significant episode with severe headaches and vertigo, a stiff neck and back and generalised muscle pain, signifying that the central nervous system has now become involved with a possible progression to viral meningitis and encephalitis*[3]*. Clinical recovery at this stage is normally possible, but does not preclude further effects of the illness in later years. It has to he remembered that ME is a life-long disability where relapse is **always** possible.

d. FINAL STAGE*[1,2]*:

After a variable interval, a multi-system syndrome may develop, involving permanent damage to skeletal or cardiac muscle and to other "end organs" such as the liver, pancreas, endocrine glands and lymphoid tissues, signifying the further development of a lengthy chronic, mainly neurological condition with evidence of metabolic dysfunction in the brain stem. Yet, stabilisation, albeit at a low level, can still be achieved by appropriate management and support. The death rate of 10% occurs almost entirely from end-organ damage within this group (mainly from cardiac or pancreatic failure). It has to be said that suicide in younger patients and in earlier stages of the disability is related to the current climate of disbelief, rejection of welfare support and loss of educational and employment prospects. It is an additional and potentially avoidable factor.

What are the Late Effects of ME?

Most doctors with substantial experience of examining these patients would agree that the outlook for any individual is unpredictable. Case records need to be kept up to date for prolonged periods because patients who have remained clinically stable over 40 years or more and have worked normally for most of their lives are still subject to significant late effects. These include: overwhelming fatigue both physical and

mental; cognitive disturbances; muscular and joint pain; muscular weakness and wasting; difficulty with breathing; episodes of hypothermia and low blood pressure; problems with swallowing and voice production as well as sudden attacks of breathlessness while sleeping. The similarities of these symptoms to those complained of by sufferers from the Post-polio syndrome, is striking and requires further explanation.

Which Group of ME Sufferers are Chiefly at Risk of the Late Effects?*[4]*

The majority of ME patients contract their illness in the 3rd and 4th decade (50%) with secondary peak at puberty (18%). The incidence at the extremes of age (below 10 years and above 50 years) has, until recently, been low (about 10% in each group). Epidemiological surveys made between 1988 and 1998, in 2 Essex hospital clinics dedicated to ME, indicate that the percentage of patients over 50 years of age attending with new illness has risen from 6% in 1988 to 16% in 1995 and 18% in 1998. Some patients indeed, present with paralysis. Others have a vague past history of illness in childhood. Some years of "weakness" or "growing pains"; recovery, but always poor at sport; possibly a stable work record for 25 years or more, followed by a decline in walking ability; unusual fatigue after simple tasks; problems with climbing stairs, dressing and with short term memory. The current age range of these patients is from 40-92 years, so it is not easy to dismiss their symptoms as due solely to "aging". Their social and medical problems are especially severe, as they share all the difficulties of access to remedial and support services complained of by other disabled people in the same age group. However, in the UK, there is almost nowhere to refer patients with suspected post-polio symptoms as the medical profession has largely forgotten or never experienced the many manifestations of that disease. Successful immunization against only 3 polio viruses among some 69 enteroviruses currently in circulation is deemed to have solved all problems!

What is the Post-Polio Syndrome?*[5,6,7]*

Poliomyelitis is an acute enteroviral infection with a wide range of clinical manifestations and multi-organ involvement (a fact which was frequently overlooked by physicians dealing with large numbers of dangerously paralyzed patients, between 1940 and 1950). 95% of people who contract the infection remain symptom free or suffer only a trivial non-specific

respiratory or gastrointestinal illness as in ME.

Some 5% of those contracting the minor illness develop muscle weakness or paralysis before more serious or fatal complications supervene. The diagnostic distinction between "paralytic" and "non-paralytic polio" was entirely arbitrary in the days of the big epidemics. In fact, the category of "non-paralytic polio" contained many patients with mild or temporary paralysis and with encephalitis, which occurs in patients reaching the later stages of this illness. Modern studies indicate that overt paralysis in these patients depends entirely on the percentage of spinal nerve cells destroyed. For damage to be visible as weakness or paralysis at least 50%-60% of the nerves controlling muscular action must be damaged or destroyed. Thus, patients with less damage who may only have had a minor illness, and some who were asymptornatic can still present many years later with a classic Post-polio syndrome.

Recent publication*[6,8]* of this information (originally derived from studies made in 1955) has resulted in a re-definition of the post-polio syndrome and will certainly include many patients currently seen in ME clinics.

Suggested New Criteria for the Diagnosis and Assessment of the Post-Polio Syndrome*[7]*.

a. A history of remote paralytic or **non-paralytic** polio, or findings on history, physical examination or laboratory and other technical studies compatible with damage to the central nervous system in earlier life.
b. A period of recovery.
c. A period of stable functioning for 10-50 years.
d. New symptoms for which no other explanation can be found. Many patients and research workers point out that the assessment of sufferers will now have to become more holistic, that standard electrical tests of muscle function (EMG) will have to be more widespread (and repeated), and that manual muscle testing must refer to repetitive activity and daily tasks rather than a single examination on the couch*[16]*.

Is it Possible that Many Patients Diagnosed as Having ME are Sufferers From an Illness Clinically Identical to "Non-Paralytic" Polio?*[6,8]*

Yes, undoubtedly! This is an important question with

fundamental implications for further research into the diagnosis, treatment and prevention of both disabilities.

Modern research published currently in a dedicated supplement of the American Journal of Physical and Medical Rehabilitation by the Editor and 3 leading research teams*[6,8]*, indicates that part of the current difficulty in obtaining a clear diagnosis of the post-polio syndrome lies in the error of dividing acute poliomyelitis into "paralytic", "non-paralytic", "abortive" and "sub clinical" categories. It has to be recognized that there is a **wide range** of nerve damage in every patient. The Post-polio syndrome may therefore include:

a. Patients whose nervous system damage was not clinically obvious at the time of diagnosis.
b. Those who had minimal paralysis for a short period and were misdiagnosed as non-paralytic polio.
c. Those patients suffering from infection due to **non** polio enteroviruses with potential to cause nervous system damage and the "Post polio" syndrome, equal to that of polio viruses e.g. Coxsackie viruses A9, A7; Coxsackie B viruses 1-6; ECHO virus 9; Enteroviruses 70, 71 - all of which have been implicated in outbreaks of ME or epidemics clinically identical to paralytic poliomyelitis.
d. Patients with symptoms clinically identical to the Post-polio syndrome whose nerve damage arises from some other cause, for example, local muscle problems due to metabolic dysfunction, the effects of persistent virus infection, immune reaction to fragments of viral genetic material etc.

It is essential that patients with clinical symptoms suggestive of Post-polio syndrome should be referred to a Physician to exclude other nervous diseases (eg, Motor Neurone Disease), and especially those which are treatable.

Is it Necessary to Differentiate Between the Late Effects of ME and the Post-Polio Syndrome?*[8,9,10]*

Not really, even if it were useful or practicable to do so at present, as the two conditions are clinically identical and similar in respect of neuroanatomical, neuroendocrine, neuropsychological electroencephalographic and other techniques, including brain imaging and molecular biology, as indicated by a remarkable series of research papers published by Bruno and colleagues over the past 20 years.

What is the Evidence That the Late Effects of ME and Post-Polio Syndrome can be Caused by Enteroviruses Other Than Polio Viruses 1-3?

a. *[11]* In 1948, the year in which polio viruses were first cultured, specimens from 2 children with clinical poliomyelitis, yielded a non-polio enterovirus, (eponymously called Coxsackie after the neighborhood in which they lived). This finding opened a Pandora's box of some 70 previously undiscovered enteroviruses of which 14 strains were later found to have neurogenic potential equal to that of polio viruses.
b. *[12]* From the late 1940s, studies in the USA indicated that outbreaks of major or minor enteroviral illness (eg. Paralytic or non-paralytic and non specific "summer 'flu") could be caused by varying proportions of virulent and non virulent polio viruses combined with other neurogenic enteroviruses, for example in Akron and Cincinnati [Table 1], Ohio (1947) Delaware and Connecticut (1949).
c. *[13]* In the UK, an outbreak of poliomyelitis affecting an Edinburgh housing estate from August 1961-February 1962 (a period when polio immunization with the Salk (injectable) vaccine had recently been introduced) provided evidence that a "mosaic" of enteroviruses, including Polio type 3, Coxsackie viruses B2 and B4, Echo viruses 5 and 15 could act in combination to enhance virulence in individual patients, to block the spread of polio virus type 3 and to interfere with vaccine efficiency. Each virus type appeared sequentially until the arrival of Echo virus 5 in November which ended the outbreak by the following February (as indicated by serial sampling of the local school sewer). It has to be remembered that a sudden change in the virulence and spread of enteroviruses in the 20th century has been due to alterations in human hygienic behavior rather than to viral mutations.

TABLE 1. 1947 OUTBREAK OF SUMMER 'FLU, CINCINNATI, USA *[12]*

DIAGNOSIS	**CASE** No	**CSF** CELL COUNT	**VIRULENCE OF** POLIO VIRUS	**3 LAB TESTS** FOR COXSACKI VIRUS(NON POLIO ENTEROVIRUS)	**SUMMARY OF** LABORATORY FINDINGS

Summer Flu	1	0	Polio HV	-	-	-	High virulence polio virus only
	2	40*	Non paralytic polio LV	-	-	-	Low virulence polio virus & infection ofCSF
	3	3	"	+	+	+	Low virulence polio virus & coxsackie virus
	4	150*	"	+	+	+	Infection of CSF & low virulence polio virus & Coxsackie virus
Non Paralytic Polio	5	734*	Polio HV	-	-	-	Only high virulence polio virus &infection of CSF
	6	27	Polio HV	-	-	-	Only high virulence polo virus
	7	70*	-	-	-	-	Only Coxsackie virus & infection of CSF
Paralytic Polio	8	107*	Polio HV	-	-	-	Only high virulence polio virus and infection of CSF
KEY * Raised cell count in cerebro-spinal fluid indicates infection in the central nervous system Polio HV - Polio type one, virulent.							

Polio LV - Possibly polio type two, low virulence (non paralytic)
Coxsackie is a neurovirulent, non polio enterovirus

How May Symptoms of the Late Effects of ME and Polio be Explained?*[9,10]*

It has to be accepted that some degree of encephalitis has occurred in all these cases and that the **areas chiefly affected include the upper spinal motor and sensory nerve roots** and **the spinal nerve networks traversing the adjacent brain stem (a nerve centre controlling all vital bodily functions which is always damaged).** The most troublesome symptoms of both conditions are **progressive muscle weakness, fatigue and pain,** and the commonest cause of relapse **over use of repaired nerve networks and an inappropriate response to physical or mental stress** in combination with the **increasing effect of normal aging.**

Fatigue: This is almost always central and due to damage affecting the Reticular activating system (which keeps the brain awake and alert as well as maintaining some control over muscular activity). Fatigue is characteristically intermittent, but profound and incapacitating and related even to minor activity.

*[14]***Muscle Weakness and Wasting:** This may have a central cause (as above) or a local origin due to loss of motor units controlling individual muscles (including the breakdown of repair to these over time). Metabolic, immune or ongoing viral injury to muscle fibres, are other possibilities where infection persists.

Pain: This is a severe symptom which is difficult to treat and is usually due to dysfunction of the thalamus, an important sensory relay station in the brain stem. Failure to produce natural painkillers (e.g. endorphins and encephalins), may be an additional factor.

Inappropriate Reaction to Physical or Mental Stress: This also arises from injury to the brain stem which normally controls the production of cortisol (a steroid required for stress control) via the hypothalamus, pituitary and adrenal glands . In the absence of an efficient response, even minor stress can cause catastrophic collapse in these patients. NB. Because of the many and varied symptoms arising from encephalitic damage to the brain, all symptoms reported, however bizarre they may

seem, must be taken as possible evidence of organic disease.

Management:

Despite promising reports from the USA of anti-enteroviral agents*[18]*, and of Dopamine receptor agonists*[9]* (to correct some deficiencies in neurotransmission) no specific medical treatment is yet available in the UK and the main principles of management rely upon **conservation of energy, reduction of stress, and simplification of manual tasks at home or at work.** These objectives cannot possibly be achieved without financial and social support, aids to mobility, house conversions and suitable rehabilitation facilities. In the USA it is claimed that (with counseling, if necessary, for those who find such adjustments to life style difficult) 91% of patients will stabilise in view of the fact that, at this stage, the disability is only slowly progressive. Patients have to be cautious about drugs, especially those acting on the central nervous system including psycho-active preparations and alcohol. In general, these patients need less anaesthetic but higher doses of pain killers than usual and more time to convalesce from surgery. There are now many new options for muscle problems including modern orthoses and corrective surgery.

Comment*[15]*

a. **There has been little government interest or support for patients suffering from the late effects of ME or from the post-polio syndrome.** It is generally expected that survivors of polio will gradually disappear because of successful immunization of the UK population 40 years ago. However the fact that "Post-polio", by any other name, can arise from **currently** circulating enteroviruses has not been taken into account. The Chief Medical Officer's Working Party on ME (set up in 1999 and funded privately by the Linbury Trust) has made it clear that its remit is **only** with management, and that all discussion about the cause, epidemiology and social benefit requirements of these patients is ruled out. It seems that it will be difficult to advise on rational management in the absence of such vital information.
b. **The potential size and cost of the problem.** This is impossible to assess in the UK because no **official** epidemiological surveys have been made. However, increasing numbers of patient support groups and individual research workers have been making their own

calculations. In the case of ME, prevalence appears to range from 300/100,000 to 500/100,000 in occupations at high risk of infection*[2]*, but no information is yet available about the number likely to suffer late effects (except that it may have trebled in the last 10 years)*[4]*.

The number likely to be affected by the post-polio syndrome has been calculated as between 200-270/100,000 currently*[7]*, but no account has been taken of survivors from non-paralytic polio which could easily double that figure. Possible costing for ME support has been based on 3 times the cost of maintenance for multiple sclerosis on the supposition that ME is 3 times as common*[4]*. The only costs that we can be **sure** of are those derived from the **failure of appropriate management**, and of **inappropriate assessments** which waste vast sums of money and medical time while allowing patients to deteriorate unnecessarily.*[16]*

c. **Some Immediate Steps that Could Be Taken***[7,17,18,19,20]*. These patients could be referred to NHS rehabilitation clinics and welfare facilities as for any other chronic neurological disease but physiotherapy must include exercise suitable for patients with some damaged muscle fibres which have been overused while others are normal and liable to deconditioning*[7]*. Separate "ME" and "Post-polio Clinics" are more expensive and often inaccessible. We should be educating doctors and paramedics **now** about the very common and seriously disabling effects of neglect*[7]*. Rapid diagnostic tests for enteroviruses, anti-enteroviral drugs and possible vaccines are already in preparation here, or in use (in the USA) to deal with the tremendous burden of circulating enteroviral infections, (for example, leading to febrile respiratory infections, viral meningitis and myocarditis, let alone unnecessary admissions to hospital and inappropriate prescription of antibiotics in children)*[17,18]*. These methods could well be employed for the benefit of young people in the UK and to prevent the rising tide of ME in schools - the commonest cause of long term absence and subsequent educational deficit!*[19,20]*

d. **Research workers must be encouraged and appropriately funded to work in this field.** However they should first be directed to papers published before 1988, the time at which all specialized experience about poliomyelitis and associated infections seem to have vanished mysteriously!*[11,12,13]*

More Woes for Top Insurance Doc

[From *Franks* Magazine - Issue 386, October 2, 2002]

The hits just keep coming for Dr. Jack Richman, Supremo of AssessMed, the medical assessment outfit more insurance companies turn to for medical opinions favourable to them kicking claimants off the disability rolls. Richman, a skeptic truly worthy of the name, has been known to opine that only three per cent of the patients he sees are actually disabled, their doctors' opinions notwithstanding. The rest are faking it. The insurance companies who hire Richman, needless to say, save loadsadough.

How high is AssessMed's refusal rate? Ask Canada Life, which was so impressed with AssessMed's track record that in February 2000, they flew one disability claimant all the way from Alberta to Toronto, all expenses paid, for a session with Dr. Richman. All of which might seem a little less than cost-effective for the insurance company. Fortunately, the good doctor made it worth their investment, declaring the patient's multiple environmental sensitivities and sleep disorders just -- quelle surprise!-- so much hypochondria. Claim denied. Canada Life, faced with subsequent medical opinions from an actual specialist in Alberta, subsequently had to pay up, but GP Richman's assessment saved them thousands in the meantime.

Readers will recall the good doctor was recently spanked for failing to ensure that one of his doctors, Dr. Hemendra Shah, was qualified to perform neuropsychological assessments.

Shah, although to this day listed as a neuropsychologist on AssessMed's website, and responsible for overruling countless opinions from qualified professionals in favour of insurance companies, is not registered with the College of Physicians and Surgeons to practice in this area. He also has on his record a two-year suspension for professional misconduct, conduct unbecoming a psychologist.

Richman, is still locked in a libel battle with the CBC's *fifth estate* for a segment on the Shah affair which he says painted him as a mercenary henchman of the insurance industry.

Further complicating matters for provincial regulators is that the

versatile Dr. Shah also does forensic psychology consultations for the Ministry of the Attorney General. Surely, though, he's registered with the College to practice in this area? Er, not as such. Wait'll the defence lawyers get ahold of this one…

Shah, although no longer authorized to do assessments on the DAC (auto accident) roster, is still doing his thing for Workplace Safety and Insurance Board (WSIB). Which begs the question: If he's not a qualified neuropsychologist as far as the DAC is concerned, why is he kosher for the WSIB?

Another regulatory decision regarding Dr. Richman is expected within weeks and, judging by the last hearing in June, it doesn't look good for our crusading doctor.

The trouble started in February 2000, when yet another motor vehicle accident victim Richman accused of malingering, Rhona DesRoches, complained to the College of Physicians and Surgeons that Richman had "performed an inadequate evaluation of her" and produced "a biased and inaccurate report dated July 22, 1998."

In its October 2001 decision, the College took issue with the wild-eyed hatchet job Richman did on DesRoches in his report: "In our view, Dr. Richman's use of such terminology as "bizarre" or "cunning" and his bald statements that Ms DesRoches' motivations were purely financial, betray a lack of understanding of the role of the objective assessor…An assessor could state, for example, that a subject showed "no evidence of pathology,". This is a far cry from Dr. Richman's choice of loaded, pejorative words such as "cunning" or "malinger."

The College recommended an oral caution, and Richman appealed. The last hearing, in June, went poorly for him. Richman arrived under protest, whining that he had already apologized twice for his monkeyshines and that attending the hearing was a chore because he was suffering from a herniated disc (*shurely not malingering?!--ed*.) Also present was a legalist from the fifth estate.

The panel was so disturbed by the evidence in the hearing that they requested the entire file of complaints against Richman. At which point Richman piped up, "All the complaints, or just those alleging bias?"

Er, just the bias complaints will do, came the reply.

But it's not all disciplinary hearings for Dr. Richman these days. Far from it. The Canadian Society of Medical Evaluators, of which Richman is secretary-treasurer and founder (Dr. Arthur Amies, his partner in AssessMed, is president), is putting on a conference in Toronto, Sept. 26-29, for their fellow medical and insurance professionals, blandly entitled "Symptoms, Diagnostic and Disability Validity: Improving Patient Outcomes."

A post-conference workshop sounds a little more up the alley of Dr. Richman, who is listed as the conference's "scientific co-chair." The workshop is called "Clinical Assessment of Malingering and Deception."

The sole "gold sponsor" of the weasel-fest? Er, the scrupulously impartial WSIB, which still accepts AssessMed's talented Dr. Shah as a qualified neuropsychologist, even if the nitpicking College doesn't.

IME Update - By: Mary Ellen, Manager - Special Projects

Thank you to all those who have filled out our Independent Medical Examination (IME) or Functional Abilities Evaluation (FAE) National Registry Submission Form and who have sent us copies of letters of complaints about their IME's to the College of Physicians and Surgeons and other regulatory bodies. Thank you also to those of you who have allowed us to pass on your name to appropriate media. We ONLY pass on your name when you give us permission! As you can see, from the article above, the media is paying attention and so too are the Colleges. With your help, the **National ME/FM Action Network** will continue to put pressure on the Colleges across the country, on the media across the country, on provincial and federal governments to work towards fair treatment of those disabled and dependent on insurance companies, CPP and WSIB for financial support. When we were healthy and able to work, we paid premiums in good faith and now, if we are unable to work, we need the peace of mind to know that our financial situation will be looked after. It is stressful enough to deal with disability and a difficult illness. Financial issues should not be added stresses.