

### **Positive results in trial of Xepol for PPS**

In February 2007 Polio Particles reported the results on a randomised, placebo controlled, double blind trial of intravenous immunoglobulin for PPS. In this type of trial participants are randomly assigned to either the group that receives the drug being investigated or the group that receives a placebo. Neither the participants nor the researcher that administers the drug/placebo know what the participant is receiving. The polio survivors who received the immunoglobulin showed some increase in muscle strength, activity levels and vitality six months after treatment while those who were given the placebo did not. Now the Swedish pharmaceutical firm Pharmalink has released the results of a similar follow-up study of 142 patients 12 months after they received the drug, which is being registered with the name Xepol. Pharmalink reports that the results *strengthen the position of this novel treatment modality for PPS by demonstrating a reduction of inflammatory cytokines in the cerebrospinal fluid and a significant reduction of symptoms of PPS while also showing that Xepol is safe and well tolerated with few or no side effects.* [Cytokines are chemical substances that enable body to communicate. They are involved in the regulation of inflammation and immunity]. The polio survivors who received Xepol showed a reduction in pain and muscle weakness and improvements in walking ability and on some measures of physical and psychological wellbeing. Xepol is described as *the first medical PPS treatment,[it] is an injectable biologic product, administered once every 9-12 months, which down-regulates the inflammatory process in the nervous system of PPS patients...Pharmalink is actively seeking a partner to bring Xepol to the market. More than 1,000 patients have been treated with the drug and many return on an annual basis for new treatment courses.* An account of the Xepol trial can be found at [www.pharmalive.com/News/index.cfm?articleid=566113&categoryid=40](http://www.pharmalive.com/News/index.cfm?articleid=566113&categoryid=40). It is not yet available in Australia.

### **Orphan diseases and orphan treatments**

Orphan diseases are defined as diseases that are so rare that there are less than 5 cases per 10,000 of the population. The majority of orphan diseases are inherited and caused by defects or mutations (changes) in genes. Post-Polio Syndrome which is listed on orphan disease website lists is of course not genetically caused. A GP in a busy practice would not expect to encounter more than one case of an orphan disease a year and may well not have learnt about the disorder during his or her training. The American FDA (Food and Drug Administration) *Consumer Magazine* says that *as disparate as rare diseases are, patients share many common frustrations. For example, for one-third of people with a rare disease an accurate diagnosis can take one to five years. And people are often so isolated that they may never know anyone else with the same disease. Patients often have to travel long distances to visit the few doctors*

*knowledgeable about their illnesses, and the costs involved with diagnosis, treatment and other related expenses can be exorbitant.*

In 1983 the USA passed the *Orphan Drug Act* to encourage the development of drugs which are needed to treat orphan diseases but would be prohibitively expensive and unprofitable to develop. Under the Act companies that develop such drugs are given tax reductions and a marketing monopoly for an extended period of time. Xepol, described above, has already received Orphan Drug Designation by the USA. In the USA medical devices are not eligible for orphan designation but in 1996 the Humanitarian Device Exemption was made to the Safe Medical Devices Act. This allows a medical device to be approved without expensive clinical trials if the manufacture can demonstrate that is safe and will probably benefit people with an extremely rare condition (affecting less than 4000 cases per year in the USA). Collaboration by researchers investigating a range of orphan diseases is also occurring. For example, the ENMC (European Neuromuscular Center) is concerned with the investigation of rare neuromuscular diseases which includes PPS. Treatment of a group of different rare diseases may have some similarities e.g. ventilatory support for respiratory insufficiency. The role of support groups, many of which operate via the Internet, is extremely important for people with orphan diseases both in providing information and social and psychological support.

### **Polio Paralympians**

Polio athletes participating at the 2008 Paralympic Games included Xiao Cuijuan from China who won a gold medal for power lifting. She contracted polio at the age of one. Heba Said Ahmed from Egypt who had polio as a child won a gold medal in power lifting having previously won gold at the Athens Paralympics. Heba broke a world record in her 181 pound class by lifting 341 pounds. She is 5 foot, 1 inch tall, walks with crutches, swinging her legs forward from the waist. Heba was written up in several US newspapers because despite her sporting successes there was little recognition of them in Egypt where on her return she continues to be treated as a second class citizen. Ironically, wrote the New York Times, *during the Olympic Games, which preceded the Paralympics, Egypt did poorly, earning only one bronze medal. But in the Paralympics, Egypt won 12 medals, including four golds. 'Face savers', read the headline on Al Ahram Weekly, an English-language Weekly newspaper. In is an extraordinary achievement coming from a country where physical disabilities are largely seen as props for street begging.*

### **Dilemma for Pakistanis opposed to polio vaccination**

Polio Particles reported two years ago that Saudi Arabia had made it mandatory for Haj pilgrims who were less than 15 years of age to provide proof of polio vaccination. Children arriving from countries where polio is endemic were given vaccination at Saudi border points. The increase in the number of cases of polio in Pakistan this year led the Saudi Arabian government to issue a directive to the Pakistani Ministry of Religious Affairs that *all* Pakistani pilgrims going to Saudi

Arabia to perform the Haj must have had at least two doses of polio vaccine, verified by a polio vaccination certificate. This occurred against a background of strong resentment and resistance by some Pakistani religious leaders to their country's national anti-polio campaign. Some have declared that polio vaccination is 'haraam' (forbidden), an un-Islamic act. They have threatened parents and forcibly driven them away from health teams administering the free vaccine. The irony is that *now the same people, many of whom are intending to perform the Haj this year, will be up against their own 'fatwas' (edicts) and would find it difficult to wriggle out of the situation because they would be required to take the same antipolio drops that they themselves have declared 'haraam'* according to Pakistan's *The International News* (26/9/2008). Many religious leaders opposed to vaccination also have relatives who plan to attend the Haj. Authorities are said to be making a database of religious leaders who have challenged the vaccination campaign that will be released to the media *when these 'elements' would kiss the holy ground* i.e. attend the Haj. Some of those who had given fatwas in the past against the anti-polio campaign [are] *now using different tactics such as issuing positive fatwas and [saying] the vaccine is now 'halal' as it is being imported from Muslim countries.* On 3/11/08 *The International News* reported that the first 'polio-free' Haj flight from Pakistan reached Jeddah on Sunday. Two drops of oral polio vaccine were administered to every passenger at Jeddah airport. *Interestingly, no less than 27 persons who spread rumours by issuing religious edicts against polio vaccine have embarked on the holy pilgrimage.*

### **Photos of polio epidemics**

Google has placed online the over ten million photos from the archives of the now defunct magazine LIFE. Many of these were never actually published. They can be found at <http://images.google.com/hosted/life> . If you type 'polio' into the search option you will find several hundred photos related to the epidemics. The captions underneath the photos relate to the associated story though these photos relating to the same story are not always placed together. There are a number for the story '*Soprano* (Australia's Marjorie Lawrence) *stricken with polio*'. Another story '*Polio mother*' shows her husband having to iron his own shirts!

### **Wife divorced over polio vaccination**

Alhaji Bulama Ali, the chief of a village in the northern Nigerian district of Maisandari, ordered his wife from their home and divorced her because she had their children vaccinated against polio. The head of the district suspended Ali from office and ordered him to reunite with his wife or face dethronement, stating that a traditional ruler was supposed to educate his subjects on the importance of immunisation. In 2003 some Islamic leaders in Nigeria condemned immunisation as a plot to make Muslims infertile. This resulted in a year's halt to immunisation in the north of the country leading to an explosion of new cases of polio. According to the BBC news (12/9/08) Ali still believes polio vaccine makes children infertile. Like many interesting stories in the press the final outcome does not appear to have been reported but in searching for it I found other stories

of Indian and Nigerian wives being divorced or threatened with divorce for having their children vaccinated or wanting to do so.

### **International post-polio awareness campaigns**

Post-Polio Health International ran a *We're still here* campaign in October 2008. Two buttons were available free from PHI, with a request that a donation be made to the organization. A red, white and blue round badge, which was intended for use in the USA, proclaimed *We're still here! One million US polio survivors, Ask me about it.* A square green button could be used in all countries and stated *We're still here! www.post-polio.org.* PHI encouraged its members to contact their local Rotary clubs with an offer to speak during *We're still here* week.

In October the British Polio Fellowship which celebrated its 75<sup>th</sup> anniversary in 2008 issued a number of press releases to increase awareness of post-polio during October. In *Nursing in practice*, a journal for primary care nurses, Graham Ball, the Fellowship's CEO cited results of a recent survey revealing that *55% of British GPs are unable to diagnose the debilitating effects of PPS. We know from members that many people who have had polio may not be aware that PPS might affect them and therefore [do] not understand their symptoms. If their GP also doesn't understand the symptoms then what chance do they have? It is clearly an urgent requirement that GPs be trained in the diagnosis of PPS.* Ball called for all cases of polio to be registered by GP practices to facilitate faster recognition of PPS symptoms. A survey of Fellowship members found that the average number of years from first going to a doctor with symptoms to receiving a diagnosis of PPS was six years.

### **Dental procedures for polio survivors**

Dr Richard Bruno has written a useful leaflet titled *Preventing complications in polio survivors undergoing dental procedures.* It can be downloaded at <http://gbppa.org/dental.htm>. It is particularly important to read and discuss with your dentist if you are undergoing dental surgery. Survivors with lung involvement are recommended to have pulmonary function tests before undergoing such surgery. Bruno notes that *breathing and swallowing can be compromised in those who had bulbar polio or chest wall paralysis, not only by anesthetics, but also merely by reclining in a dental chair. Polio survivors often have difficulty breathing or swallowing saliva when reclining. A comfortable reclined position must be identified before the procedure begins. And the procedure may need to be interrupted frequently to allow the patient to breathe fully and to swallow. Also a number of polio survivors have experienced severe neck or back pain following lengthy procedures, since their muscles spasm easily when placed in unusual or awkward positions, including hypertension (extreme bending backward) of the neck.* Getting onto and off the dental chair can be difficult and survivors may need to request help in transferring particularly if they are still partly anesthetized. Survivors typically don't ask for help. Bruno suggests avoiding long explanations. A simple explanation such as, *'My legs*

*(arms) are paralyzed and I can't get into/out of the chair. I will need help' should suffice.* If professionals make comments such as *'I bet you can do it if you tried'* you may need to repeat your comment that you need assistance. Bruno reiterates the rules about anaesthetics for polios. They typically need more local anaesthetics but less for general anaesthetics than do their able-bodied counterparts.