The Post-Polio Syndrome: An Orphan Disease

Peter Nolan FRACP
Continuing Medical Education
“I knew Jesus before he was a Superstar”

A song by Francis Henry
Beware of the Marketing Manager!
A personal history

- My Grandfather was an orphan.
  - No birth certificate
  - His mother died when aged 12.
  - His Mother buried in an unmarked grave.
  - Left school age 10 to be a house boy for the landed gentry
  - Joined the AIF in 1915 with no next of kin on his enrolment papers
  - No personal possessions when he left in 1915 and arrived back in Australia in 1919.
What is an Orphan?

- Form the Greek word *Orphanus* meaning bereaved.
- Bereaved; Rob, dispossess; usually of immaterial things e.g. life, hope etc.
  - Leave desolate.
- Bereaved of parent.
- Bereft of previous protection and advantages.
Why is Post-Polio an Orphan disease?

- No next-of-kin. Indeed some people consider that it doesn’t even exist.
- Dispossessed of any formal recognition by the modern health care system.
Sleep Apnea Syndrome after Poliomyelitis

Introduction
A variety of neurologic diseases can cause abnormal breathing during sleep, but poliomyelitis appears unique in that sleep-disordered breathing (SDB) can occur many years after apparent recovery from the acute disease. We recently studied a young man whose case illustrates several mechanisms pertaining to the pathogenesis of this delayed form of the sleep apnea syndrome.

SUMMARY We present a case of severe breathing abnormality during sleep in a young man who had had poliomyelitis 20 yr before. His sleep disorder led to respiratory failure and cor pulmonale, which were greatly improved by oxygen therapy. A study of this case and those previously described supports the notion that brainstem damage during acute poliomyelitis is important in the later appearance of sleep-disordered breathing. In addition, such patients usually have mechanical abnormalities involving the thoracic cage and respiratory muscles. These ventilatory restrictions amplify the pathophysiologic effects of abnormal central nervous system control of breathing during sleep, and we suggest that their presence has a key role in the development of sleep apnea syndrome in these patients.

AM REV RESPIR DIS 1983; 127:129-133
Respiratory function during wakefulness and sleep among survivors of respiratory and non-respiratory poliomyelitis. T.E. Dolmage, M.A. Avendano, R.S. Goldstein.

ABSTRACT: The purpose of this study was to determine whether there is a difference in respiratory mechanics and gas exchange between polio survivors and healthy, age-matched controls during wakefulness and sleep.

Polio survivors were divided into four groups. The first group included those who had evidence of respiratory muscle involvement originally (P_{RM}) and the second group included those who had bulbar muscle involvement originally (P_{BM}). The third and fourth groups had only limb involvement originally but were separated by absence (P_{SL}) or presence of a scoliosis (P_{SS}) at the time of their evaluation.

Each subject completed baseline and one year follow-up measurements of lung volumes, diffusion, flow rates, respiratory muscle strength, central and peripheral chemoreflexes and arterial blood gases. Sleep measurements included a full respiratory polysomnographic study.

Fifty polio survivors and 13 controls completed the study. The P_{RM} and P_{SS} groups had an elevated arterial carbon dioxide tension (P_{aco2}) (mean±SE 6.0±0.4 and 6.0±0.3 kPa, respectively), reduced vital capacity (2.8±0.3 and 2.9±0.3 l, respectively), reduced maximal inspiratory pressure (-5.9±0.7 and -5.4±0.8 kPa, respectively) and reduced maximal expiratory pressure (9.8±1.1 and 9.1±1.2 kPa, respectively), when compared with non-polio controls. During sleep P_{RM} and P_{SS} groups experienced a higher P_{aco2} (6.5±0.5 and 6.7±0.4 kPa, respectively) and a lower arterial oxygen saturation (S_{ao2}) (89±4 and 86±3%, respectively). There were no differences among groups for diffusion, flow rates and chemoreflexes. All other polio survivors showed essentially normal respiratory function.

A respiratory evaluation is important in polio survivors if there is a clear history of initial respiratory muscle involvement or the finding of a kyphoscoliosis many years later. This study does not support previous reports suggesting respiratory dysfunction may occur among those who had only limb involvement at the time of their acute polio.

Eur Respir J., 1992, 5, 864–870
eutic interventions. A typical evaluation consisted of seven components:

1) A detailed medical history with special attention to current health problems as well as the extent of initial involvement from polio, the period of recovery, the duration of neurologic and functional stability, and the time of onset of new health problems;

2) A physical exam with special emphasis on assessing the musculoskeletal and nervous systems;

3) An electromyogram (EMG) and nerve conduction study of selected muscles and nerves;

4) A biomechanical and functional evaluation with special attention to orthotic and adaptive equipment needs;

5) A basic panel of screening laboratory tests that were supplemented with additional x rays and laboratory studies as indicated by the history and physical examination;

6) A psychosocial assessment; and

7) Referral to other members of the rehabilitation team or other medical specialists as needed.

Based on the evaluation findings, the patient (in item 1) history of...
Natural History of Poliomyelitis of Clinic Group

% Max Function

Years

A Birth
B Onset of Polio
C Maximum Neurologic and Functional Recovery
D Onset of New Health Problems
E Time of Evaluation
F Death
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Virologically confirmed Type 1 Poliovirus epidemics
An iron-lung ward in the early days of treating polio.
Heroine's lonely grave
her only Qld memorial

RECOGNITION was at the end of a long, hard road in Sister Elizabeth Kenny's life so she should not be greatly disappointed that 40 years after her death no suitable hometown memorial exists to this international figure of the dark age of eugenics.

If you are middle aged, you still remember polio, that paralyzing disease that was struck down the 1940's and 50's. It has been forgotten since the miracle of the Salk vaccine conquered it.

But the memories remain as does SISTER KENNY's grave on a silent, rolling hill just outside her native Dunns hometown of Cobbitty.

It is not a prominent one, as Cobbitty is a tiny town, and as you approach you expect to be hardly noticed by a headstone or monument of some significance, certainly some evidence of the work of a woman who had lived through a country's history.

Sister Kenny's grave is back in the middle of a country cemetery on the side of a lonely hill.

Instead, the last earthly remains of Sister Kenny lie beneath a white slab marked "Sister Kenny, 69, 1927-1969." It is a quiet corner but disappointing, more so because the simple inscription to this international figure is not more prominent. Certainly more significant than a humble headstone.

In 1969, Sister Kenny was laid to rest in New York's St. Luke's Hospital. Her death was a tragic one, killed in a car crash.

She was a remarkable woman, who devoted her life to the fight against polio, which had ravaged the world in the 1940's and 50's.

Sister Kenny's methods of treating polio were adopted in 14 countries, and she is remembered in her hometown of Cobbitty as a miracle worker and a wonderful person.

But her legacy lives on, not just in Cobbitty, but throughout the world, where her methods have saved countless lives. She is a true hero, and her memory will live on forever.