British study sheds some light on cause of death for people with OI

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The OI Foundation’s Information on Demand service often receives questions about the most frequent cause of death for children and adults who have OI. While never an easy subject to talk about, being aware of risks helps people make choices and not just react to events. One of the OI Foundation’s goals is to give people the information they need to reduce risk and enhance life and health.

There is only one published study specifically on this topic: “Causes of Death in Osteogenesis Imperfecta,” by Dr. Colin R. Paterson and Dr. Susan J. McAllion. It was published in 1996 in the Journal of Clinical Pathology. Starting with information from a survey of 1,297 people diagnosed with OI and living in Great Britain between 1980 and 1995, 79 death reports (not counting infants with Type II OI) were analyzed. This is a small study, so it has limitations, but the information is thought provoking.

Two important trends are evident from this report:
· Respiratory failure causes a significant number of deaths among people with OI at all ages.
· Accidents that would be relatively minor incidents for others, can lead to death in people with OI.

In 1995, respiratory failure was responsible for 15% of all deaths in Great Britain. According to this study, respiratory failure was responsible for 81.6% of all reported deaths for people with Type III OI and 39% of all deaths for those with Type I and Type IV OI. Respiratory failure was the single most common cause of death for children under age 10 with Type III OI and for adults with all types of OI.

Accidental trauma was reported as the cause of death for only 2.7% of people in Great Britain at the time of this study, but it caused 13.2% of deaths for people with OI Type III and 2.4% of deaths for those with Type I and Type IV. All of the reports mentioned in the study involved falls or car accidents with multiple injuries, internal bleeding and in two cases, head injuries.

This study found that the incidence of cardiovascular disease was slightly less in those with OI than in the comparison population. Deaths from neurological problems were higher and in this study all were caused by basilar invagination. Cancer was included in a broad “other” category, and was significantly less than the comparison population but may have been under reported.

No one can control all risk and we are frequently reminded that death is indeed a part of life. However, finding ways to protect your health and learning to make decisions that promote good health can reduce your risk.

Preparation is key to eliminating stress and hardship

Recently, national media attention has prompted a number of questions from the OI community about living wills or advanced medical directives.

The OI Foundation strongly recommends you talk to your family, healthcare providers, and legal experts about your wishes—whether or not you have OI. While it is not a pleasant subject, being prepared may help alleviate stress and hardship for the loved ones you leave behind.

Advance Medical Directives provide a guideline for your caretakers and medical professionals if you are incapacitated and cannot make your own medical decisions. There are several types of documents that can be used including: Advance Directive, Durable Power of Attorney for Health Care Decision (DPAHCD), Do Not Resuscitate Orders (DNR), and Living Wills.

The following websites will help you learn about the differences between the documents and the legal requirements for each. They also contain links to free downloadable forms:

www.familydoctor.org
from the American Academy of Family Physicians, (800) 274-2237

www.abanet.org/aging/home
from the American Bar Association, (202) 662-8690

www.nhpc.org
from the National Hospice and Palliative Care Organization, (800) 658-8898
What does this mean and what can you do?

Make protecting your respiratory system a priority.
- Promptly seek care for all respiratory infections.
- Never smoke and never expose children with OI to second hand smoke. Dr. Jay Shapiro, M.D., of the Kennedy Krieger Institute in Baltimore, Maryland states that smoking should be considered lethal for all people who have OI.
- Have pulmonary (lung) function evaluated yearly by your primary care doctor or by a specialist called a pulmonologist. Children, and especially, adults who have Type III OI, and everyone with scoliosis or kyphoscoliosis, needs to know if they are getting enough oxygen from each breath. Anyone with a compromised pulmonary function test should consider a sleep study to rule out sleep apnea. Sleep apnea (when a person stops breathing for brief periods of time during sleep) can affect pulmonary function.
- Talk with your doctor and physical therapist about safe and appropriate ways to improve your lung capacity. There is a wide range of available options including deep breathing exercises, activities such as blowing ping-pong balls across a table or playing a wind musical instrument, and swimming or other vigorous exercise.
- Those with scoliosis (see the Q & A on page 6) need to monitor their spine curve for change and discuss treatment options with a qualified orthopedist.

Take reasonable precautions to avoid accidents.
- Safety experts remind us that many “accidents” can be prevented:
  - Prevent falls by providing lights in halls and stairways.
  - Put grab bars in bathrooms.
  - Store items where they are easy to reach.
  - Remove throw rugs.
- Be alert to surroundings when using sidewalks or in crowds.
- Use appropriate seat belts in wheelchairs and in cars.

Parents turn grief into action to honor son

Michael John Shultz was a cheerful baby. “No matter how much pain he was in, he could always give you a smile,” his mother Beth remembers.

Michael was born to John and Beth Shultz in August 1998 with a severe form of OI. “Michael would never break just one bone,” Beth explained. “He would break multiple bones at a time.”

Like other parents of a child with severe OI, John and Beth knew the heart-breaking frustration of not being able to prevent the fractures from happening. Sadly, Michael passed away when he was just eight months old.

John and Beth decided that they didn’t want any child to go through the experience that Michael had. Since John was an avid golfer, the couple decided to hold a golf tournament to raise money for OI research.

The first Miracle Michael Fund Charity Golf Tournament was held in 1999, on the day that would have been Michael’s first birthday. Annual events have been held since, and together, they have brought in almost $100,000.

John and Beth have set their goal much higher. “We have a goal in our lifetime . . . we want to see a cure,” Beth said. “We also have a goal of raising at least $1 million for OI research.”

The OI Fdn expresses its sincerest condolences to all those in our community who have lost a loved one.

It is our hope that this information helps eliminate fear and provides tools for a longer, healthier life.