



The NZ Organisation for Rare Disorders

John Forman – Executive Director

Managed Clinical Networks

- An opportunity to improve clinical care for those affected by rare disorders
- Next steps

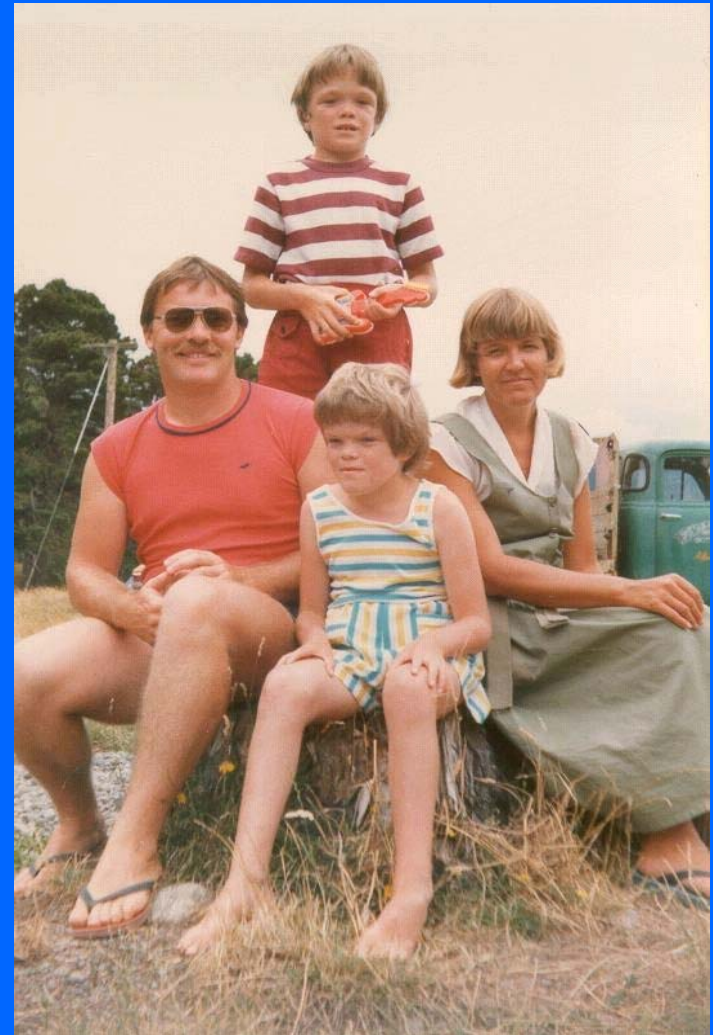
John, Judith, Timothy and Hollie

an introduction



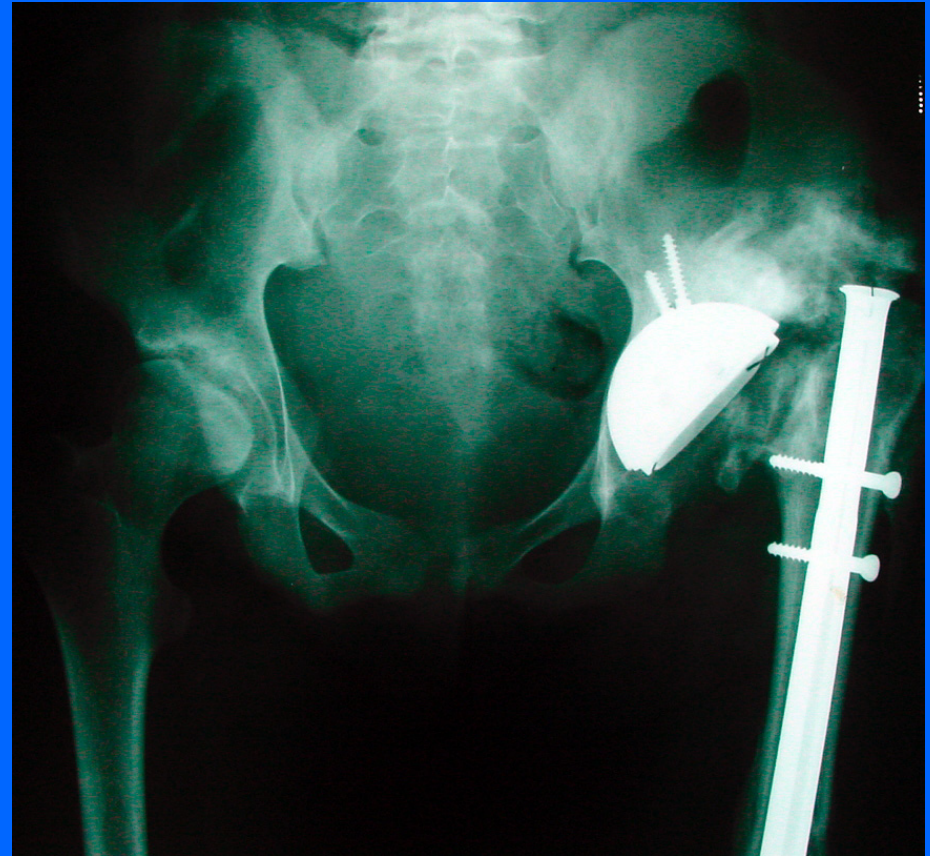
Key issues in the early years

- Dealing with hearing loss, ENT & dental problems, & moderate chronic health issues
- Intellectual disability
- Family support
- Inclusion in society: kindergarten, schools, sport, girls' brigade



Teenage to adult years

- Failed hip replacement
- Ataxia
- Psychosis
- GI problems
- Joint inflammation
- And more



Adult years

- Significant health and disability issues
- Good knowledge of genetic cause, progression and symptom management



Alpha-Mannosidosis

- One of 45 Lysosomal Storage Diseases
- Single gene defect at 19cen-q12
- Autosomal recessive inheritance
- Lysosomal Alpha-D-Mannosidase deficiency
- 1 in 1 million live births
- Storage of un-degraded oligosaccharide sugars in the Lysosome
- Insidiously progressive

That is what got me involved in:

- LDNZ – Lysosomal Diseases New Zealand
 - To improve outcomes for those with Lysosomal diseases
- NZORD – The NZ Organisation for Rare Disorders
 - To improve outcomes for families affected by all rare disorders

The common problems of rare disorders

- Lack of priority in service planning
 - Can't compete with cancer, heart disease diabetes and surgery waiting lists
- People do not live where the expertise is
 - DHBs work in geographical areas and are reluctant to cross the boundaries
 - Specialists have difficulty reaching out to those who need their care (and some professionals reluctant to refer to the experts)

The common problems of rare disorders

- Care sometimes very good for children
 - but the children grow up
 - Adult services don't deal well with many rare disorders
- Post code service delivery
- A state of relative neglect and poorer outcomes than should be acceptable

Managed Clinical Networks

Wales:

- Develop high quality, equitable and sustainable specialised children's health services across Wales based upon the best possible evidence and with the children and their carers at the centre of all planning and provision.
- The project is developing service specific standards for an identified group of 22 specialised services for children in Wales.

England

- National commissioning of specialist services

Managed Clinical Networks

Victoria

- “Clinical networks have been established so that the knowledge, wisdom and experience of clinicians is used to better plan and deliver more responsive, effective and efficient services across Victoria.”

Scotland

- “Strengthening the role of managed clinical networks”

Health & Disability Commissioner

- Lysosomal Diseases NZ – May 2007
- Complaint about services to 14/92 families
- Proposed a specialist service to manage patients
- Supported by Commissioner in report to Director-General of Health
- Accepted in principle by Ministry – March 2008

Health Minister Tony Ryall

21 October 2009

- "Cabinet has agreed to a number of proposals from the Ministerial Review Group's report 'Meeting the Challenge' that will greatly improve national and regional cooperation and reduce duplication of back office functions"
- "The Government wants better coordination between District Health Boards (DHBs) and the Ministry of Health, and we want neighbouring DHBs working better together to improve services. Clinical networks will be a big part of this cooperation."

Health Minister Tony Ryall

- The major changes include setting up a new National Health Board (NHB) within the Ministry of Health.
- The new NHB will manage national planning and funding of all IT, workforce planning and capital investment. It will also take national responsibility for vulnerable health services such as paediatric oncology.
 - Genetic services, and others are also mentioned

Next steps

- Contract from Ministry of Health to Paediatric Society of NZ
 - Development of Managed Clinical Networks
- NZORD workshop on Tuesday 17th November
 - Design of Managed Clinical Networks

Workshop Participants

- NZORD, LDNZ, Fragile X Trust, Muscular Dystrophy Association, DEBRA NZ, Rett NZ, Cystic Fibrosis Association, and more.
- A chance to get out of the “too small, too hard” basket
- We will still have to push hard to ensure we do not slip under the radar



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