HEREDITARY ANGIOEDEMA

HOSSAIN KHUNDKER

WHANGAREI HOSPITAL
NEW ZEALAND
BACKGROUND

- Mr K, 33 year male of mixed ethnicity, lives in a farm at Dargaville, Northland.
Dargville is known as the Kumara Capital of New Zealand
MR K IS ALSO A SINGER
Presented with loss of voice last year at Dargaville Hospital.
Nasal Polyps identified.

Suggested for ENT surgery

Referred to Whangarei Hospital for Immunology assessment.
Whangarei Base Hospital
Known Hereditary Angioedema (HAE) patient.

First experienced symptoms at the age of 21.

With abdominal pain and swelling.

Tested and identified HAE at that time.
Patient’s family history

- Mother
- Maternal Grand Mother and
- One of the brother of Maternal Grand Mother is known HAE patient
Hereditary Angioedema (HAE)

- Very rare and potentially life-threatening genetic condition
- About 1 in 10,000 to 1 in 50,000 people.
- Genetic defect passed on in families.
Symptoms of HAE

Episodes of edema in various body parts including the hands, feet, face and airway.
Swelling of Hand

Normal

Edema attack
Swelling of Feet
Abdominal Swelling

Abdominal Swelling Normal Abdomen
Symptoms of HAE (Continued)

- Severe abdominal pain
- Nausea and vomiting caused by edema in the intestinal wall.
Swelling of Airways

- Airway edema - risk of death by suffocation.

- Even slightest hint of throat edema should seek immediate treatment
Swelling of Airways

- Uvula
- Left Tonsil
- Lingual Thyroid
- Tongue
HAE Oropharyngeal/Laryngeal Attacks
Causes of HAE

- Defect in the gene that controls a blood protein - C1 Inhibitor.

- As a result - inadequate or non-functioning C1-Inhibitor protein.
Patient’s clinical information

- Mr K had a number of episodes with hands, lips and gums affected.
- Five years ago after some rough dental work with abdominal attacks.
- Then commenced on Donazol, continued over the years.
Pt.'s clinical info (cont.)

- Quiet asymptomatic while on Danazol.
- Never throat involvement
- Never required C1 inhibitor (Berinert) infusion.
Pt.'s clinical info (cont.)

- Scheduled to undergo ENT surgery
- Need infusion 1000 U C1 inhibitor
- Given within 2-3 Hr prior to surgery
- He is only known patient in Northland Region
Immunology Consultant suggested keeping 1000 U C1 inhibitor (Berinert) infusion in our Blood Bank all the time.
THANKS NZBS TEAM

SAVE LIVES
GIVE BLOOD
THANKS

CSL Biotherapies

Berinert™
C1 Esterase Inhibitor, Human

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Part of Our Blood Bank Team
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