

Infection Control & Prevention Practitioner & The Infectious

Snotty Nosed Kid to Less Snotty
Nosed 55 yr old
with Anderson-Fabry's Disease



**INFECTION
CONTROL
CENTER**



**NOW
HIRING**

SIGN
IN

I SHOULD HAVE SAID "I'M
NOT AFRAID OF HARD
WORK" RATHER
THAN "I DON'T
MIND GETTING
MY HANDS
DIRTY."

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10-5
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Outline

1. Describe my professional experience 10 min
 1. CV
 2. Change theory & pt/staff compliance with Nosocomial Infection Prevention & Control Precautions

2. Describe my Medical Conditions 20 min
 1. Snotty Nosed Kid & Adult
 2. Fabry's Disease
 3. The effects of IPC Precautions on my life.

3. Questions 15 min

Sheila Kennedy CV

- IPCP, Shaughnessy Hospital, Vancouver in 1991-93
- CNM-IPC unit, Riyadh Al Kharj Armed Forces Hospital, Riyadh, Kingdom of Saudi Arabia 1994-97
- IPCP, GF Strong and Pearson, Vancouver 1997-98
- IPCP, Prince Sultan Cardiac Centre, Riyadh 1998-99
- IPCP, St Paul's, Vancouver 99-2000
- Manager, IPCU, Providence Alaska, Anchorage, Alaska 2000-02
- OCHS RN-Infection Diseases only, Alaska Regional Hospital, Anchorage 2002-03
- Interior health 2003-06
- Long Term Disability



Why am I smiling? I arrived in February & hadn't experienced the singular joy of wearing head to toe black polyester in 40 + degree Celsius!!!

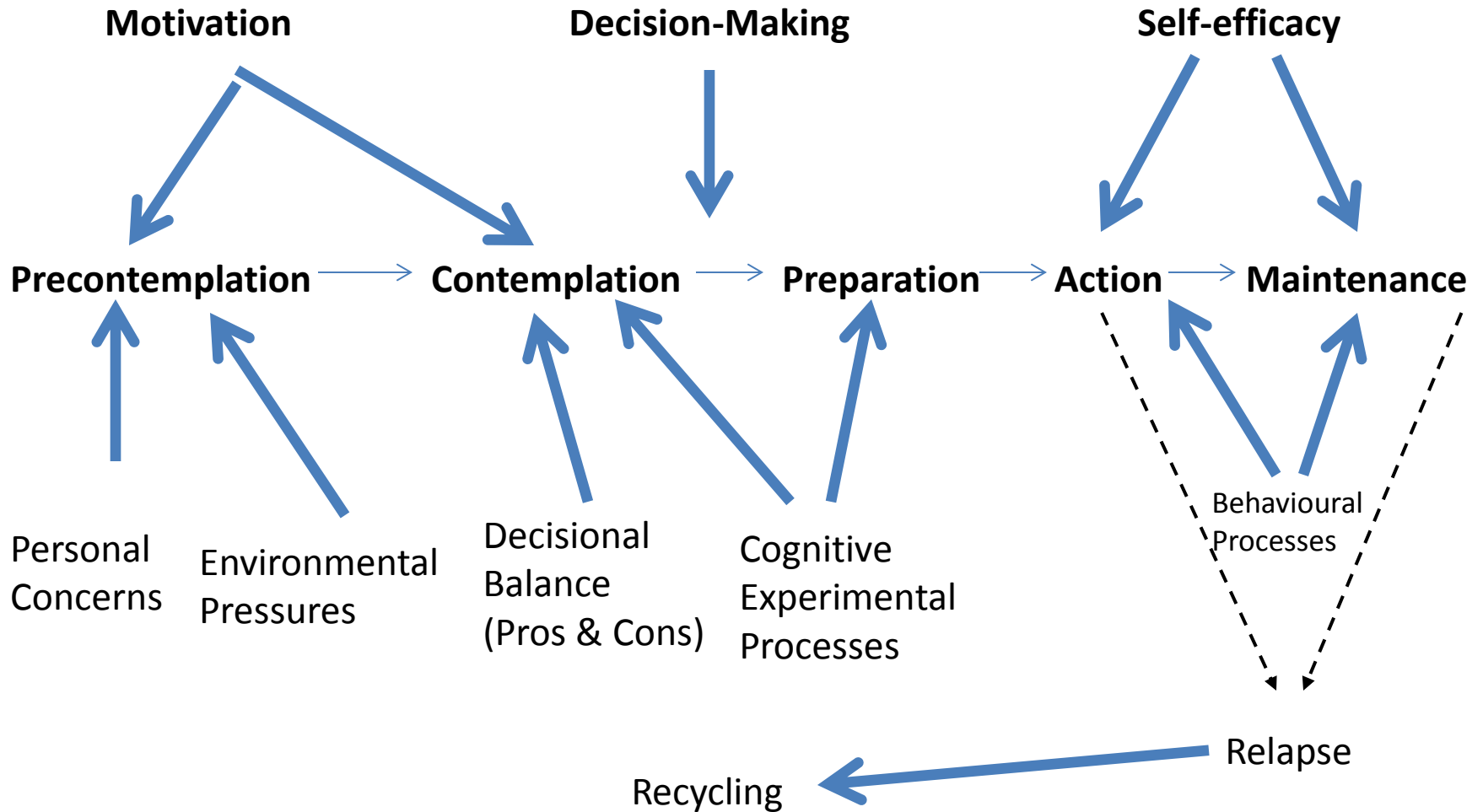
How Do People Change?

- People change voluntarily only when they
 - Become *concerned* about the need for change
 - Become *convinced* that the change is in their best interests or will benefit them more than cost them
 - Organize a *plan of action* that they are *committed* to implementing
 - *Take the actions* that are necessary to make the change and sustain the change

Stage of Change Tasks

Precontemplation	Awareness, Concern, Confidence
Contemplation	Risk-Reward Analysis & Decision making
Preparation	Commitment & Creating an Effective/Acceptable Plan
Action	Adequate Implementation of Plan and Revising as Needed
Maintenance	Integration into Lifestyle

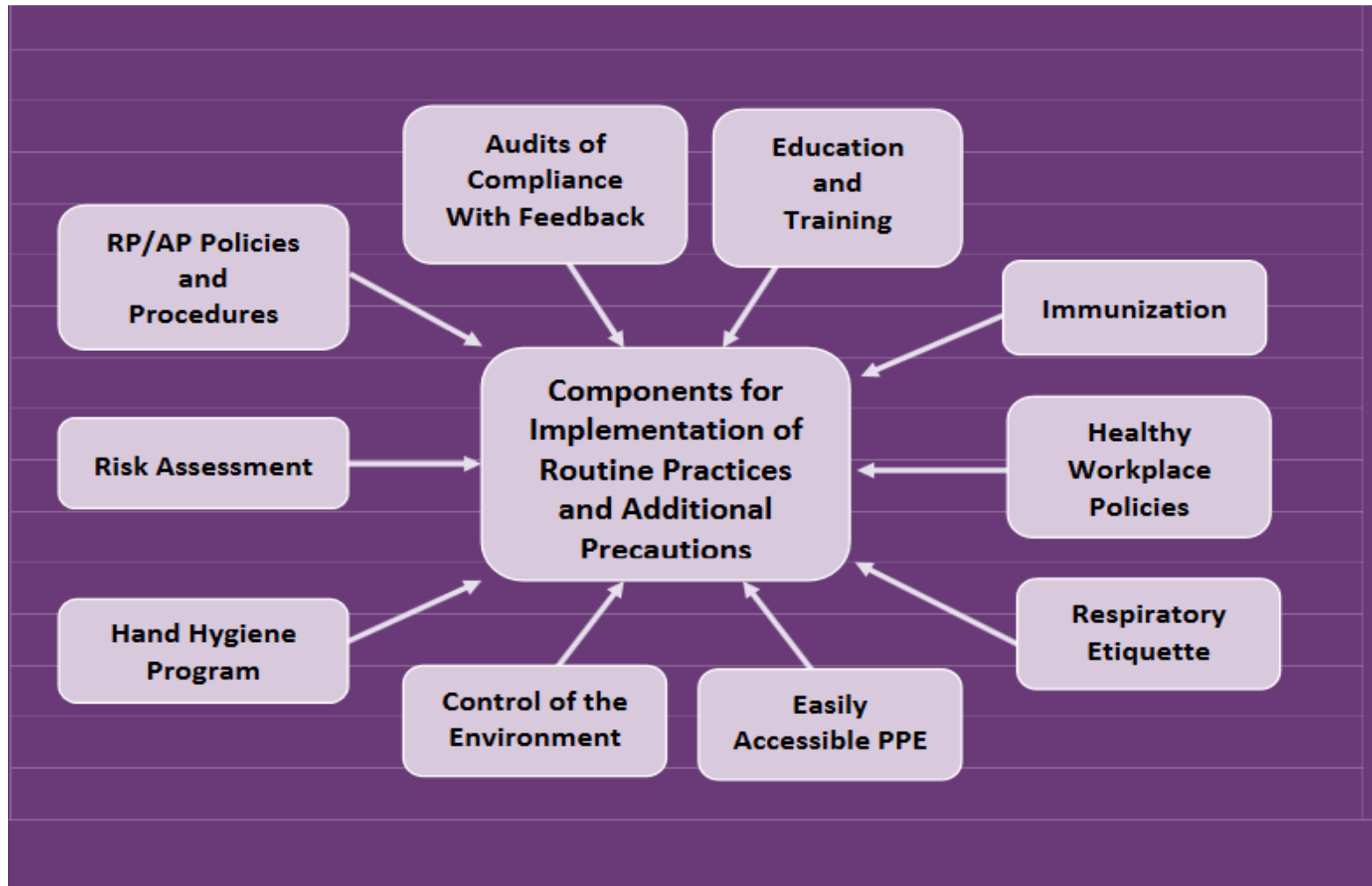
Theoretical & practical considerations related to movement through the Stages of Change





“What if we don’t change at all ...
and something magical just happens?”

Routine and Additional Precautions



Snotty nosed kid to Snotty nose adult

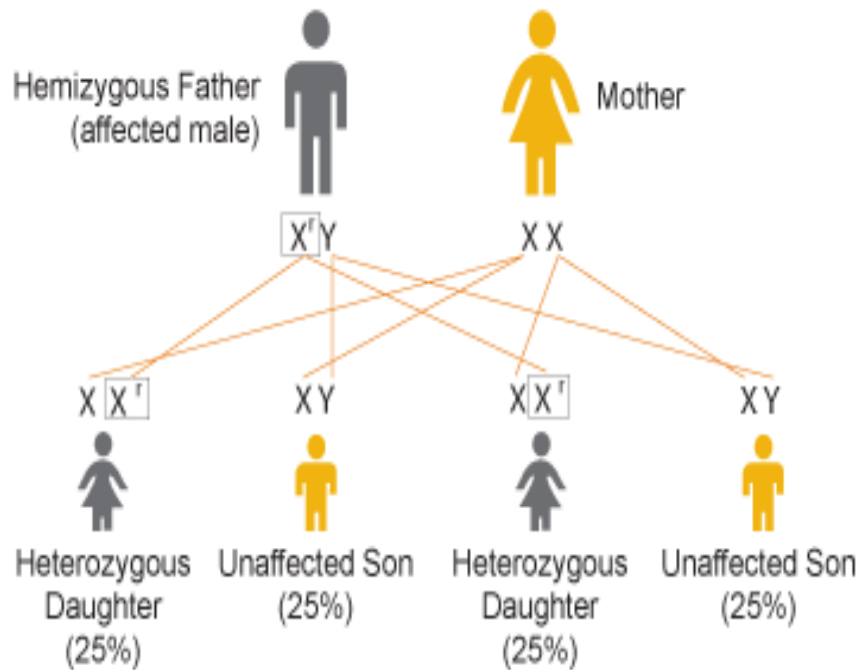
- Wide allergy pattern
- Chronic ENT infections
- Hx of Febrile Convulsions bw 2-6 yr (rubbing EHOH & ice bath horrors)
- Asthma (22 yrs)
- URTI infections tx with antibiotics >30 courses (stopped counting 1.5 decades ago)
- New antihistamines, nasal sprays etc & more ways to avoid allergens
- Basic infection control measures such as:
 - Good handwashing,
 - Avoiding contact with people who have active infections
 - Immunizations
 - Nasal lavage

Anderson-Fabry Disease

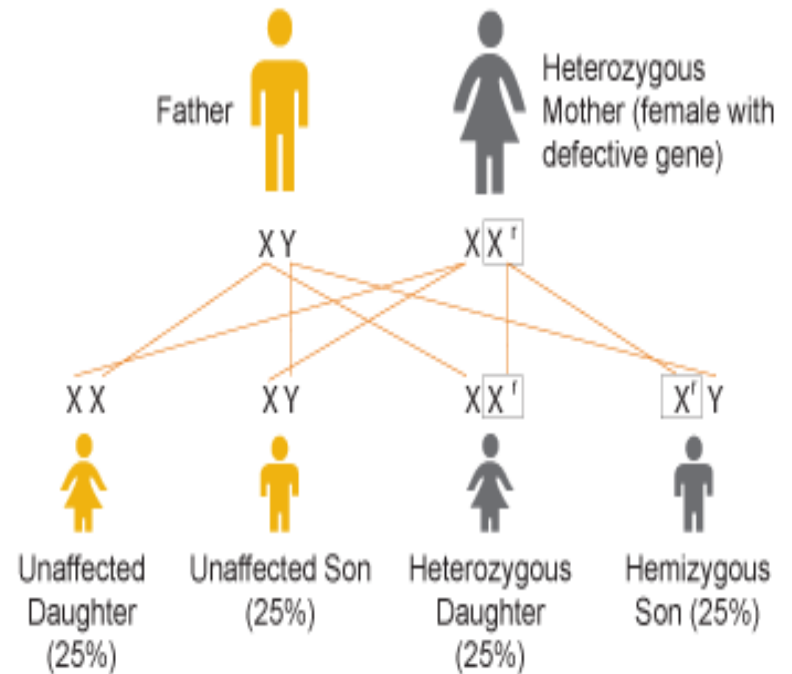
- Anderson-Fabry (europe) Fabry Disease (n. america)
- Fabry patients are missing or lack sufficient α -galactosidase A (α -Gal A) enzyme which results in sugars and fatty acids (Gb3) accumulating in the lysosomes, which function as the cells' recycling centres, and impairs the function of several major organs.
- This can become a major problem in parts of the body that depend on small blood vessels, since the build-up – or “storage” – of Gb3 can clog these vessels
- Fabry disease is a X-linked inherited disorder
- An estimated 1/40,000 to 60,000 people have Fabry disease (low due to delay/difficulty with Dx)
- Nova Scotia (1 million) has as many Fabry pts as Spain (40 million)
 - Fabry testing is recommended for **all people in Nova Scotia**

Fabry Disease Genetics

Segregation of X-Linked Trait (Hemizygous Father)

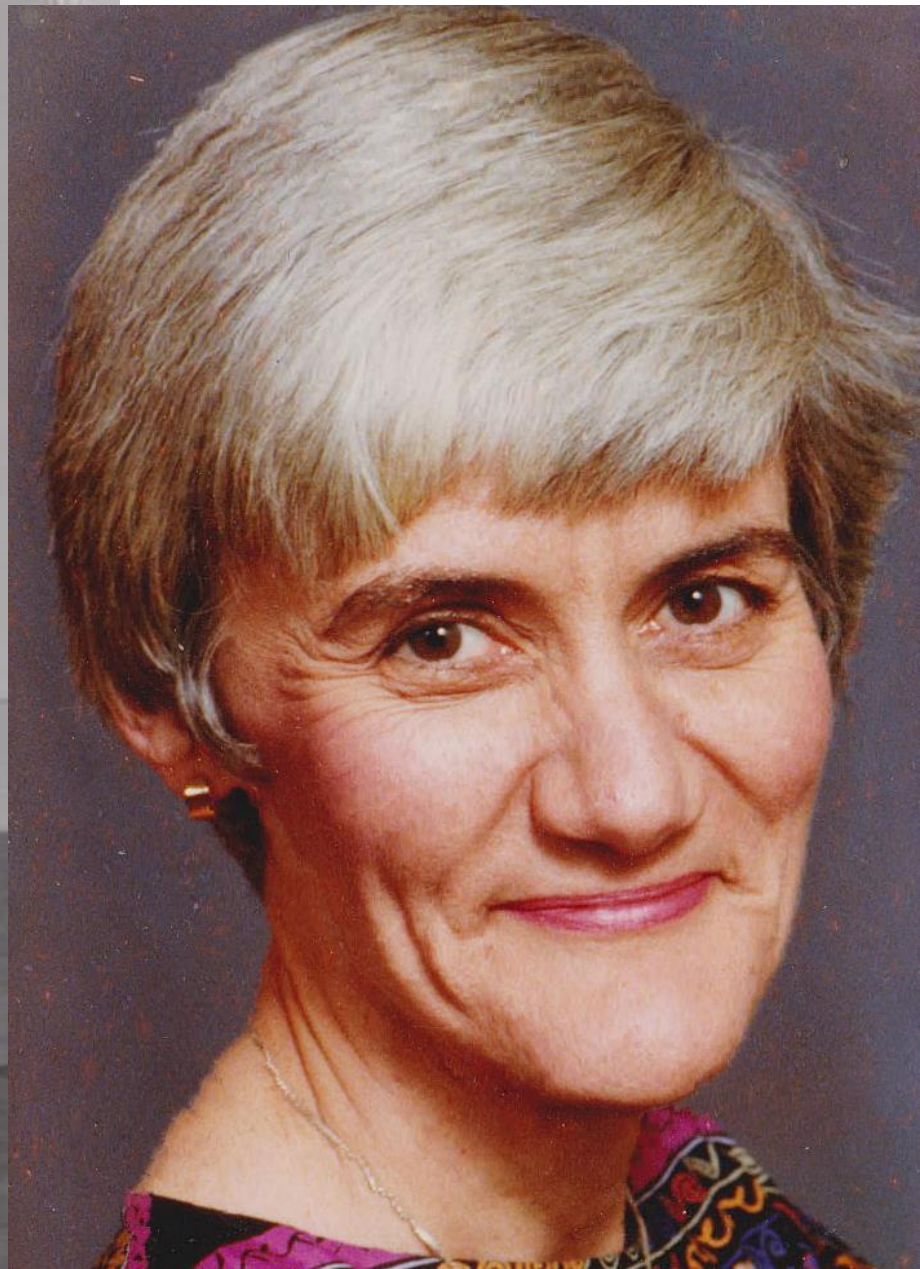


Segregation of X-Linked Trait (Heterozygous Mother)



Genetic Counselling

- I was diagnosed 1st in Spring 2003
- My Sister and Mother were diagnosed later that year
- My Mother had 2 half sisters:
 1. Aunt # 1 was deceased, so I notified her 2 adult children in England & gratefully they were negative.
 2. Aunt # 2 refused to be tested and refused to notify her adult children that they should be tested.



Fabry Disease – Systems/Symptoms

Pain	Brain	Heart	Kidney	Skin	GI	Eyes	Ears	Fatigue
Arcoparesis	TIA/Strokes	Changes in Heart wall thickness -LVH	Proteinuria	Angio-keratomas bt nipples & knees	GERD	Corneal changes – •Corneal whorls •Cataracts	Tinnitus	+++++!!!!
Arthragias/ Myalgial	Depression /Anxiety-? Primary or Secondary	Rhythm changes- Afib, Brady, LVH	↓ GFR	↓ Sweating	IBS	Sudden blindness	Hearing loss	
Pain Crises		High BP Value issues Heart attacks Heart failure	End Stage Renal Disease	Unable to tolerate extreme temp changes			Balance issues	
Triggers - sudden changes in internal or external temp , in relative humidity, & stress		Elevated Troponin						

Fabry Disease – Systems/Symptoms

Lungs	Bones	Growth	GU	Blood	Immune
SOB	↑ Osteoporosis	Delays	Infertility (males)	↓ Ferritin ↓ Hemoglobin	Immune response to GB3 in tissues
Chronic Bronchitis					

Fabry Tx

- **IV Enzyme Replacement Therapy (ERT)**

- 2 drugs available
- Estimated yearly drug cost:
>\$250,000

- **Oral Chaperone Therapy**

- Only for certain mutations (yeah me)
- Should be approved by provinces by the end 2018

Gene Therapy

The first Fabry man to receive Gene Therapy in a Phase I clinical study was in 2017 in Canada. Studies are now being enrolled to do further investigation. (donated blood marrow)

2 Hospitalizations

1. acute bronchitis

1. Tx: Prednisone/Clarithromycin-(should be used with caution in pt with heart arrhythmias)
2. Day 2 of antibiotic – 2 hours of palpitations @ 0100-0300
3. Returned to walkin clinic-sent to Vernon Jubilee Hospital on a Saturday morning
4. Admitted query **heart attack** to CCU (↑Treponin)
5. Infection Control Concerns:
 1. Inconsistent hand cleaning all CCU staff
 2. I asked if they wished to swab me for resistant bacterial pathogens due to the # of antibiotics I have ingested in my life – staff told me it was unnecessary
 3. Equipment cleaning was excellent
 4. Very little hand hygiene in radiology etc
 5. I was hesitant to asking staff if they would mind using the hand sanitizer in front of me.

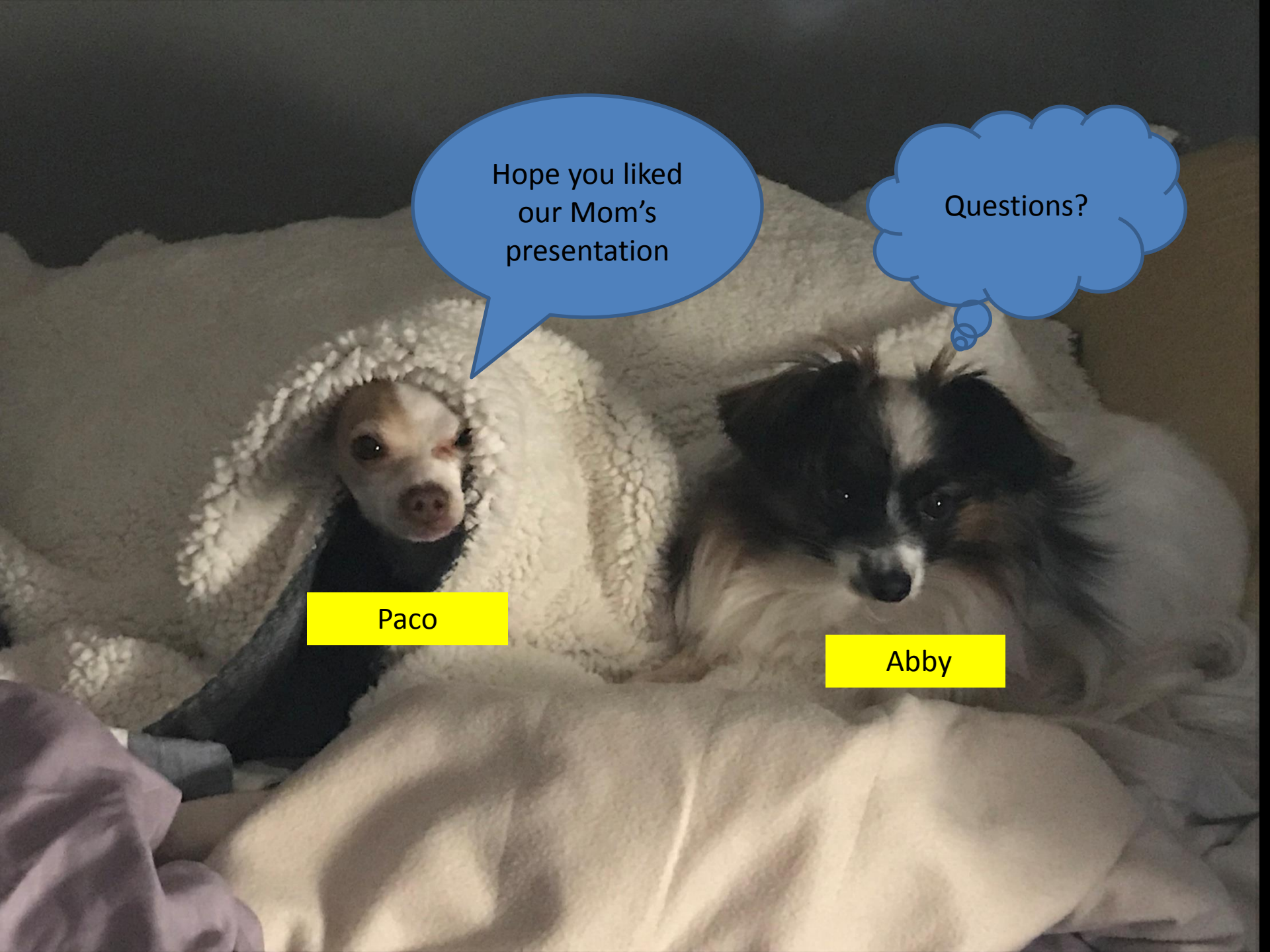
2 Hospitalizations (con't)

2. ? Syncope

1. Renal disease tx focuses on ACE/ARB, low sodium diet, weight management to lower BP
2. My family has paradoxically low BP (few readings of hypertensive - except in hospital) – +++ orthostatic hypotension
3. Bradycardia (resting pulse then was 30-50 range)
4. 2 bed room (by door) on medicine
5. Infection Control Concerns
 1. Inconsistent hand cleaning all staff
 2. I asked if they wished to swab me for resistant bacterial pathogens due to the # of antibiotics I have ingested in my life – staff told me it was unnecessary
 3. Equipment cleaning hit & miss
 4. Very little hand hygiene in radiology etc
 5. I was hesitant to asking staff if they would mind using the hand sanitizer in front of me.
6. Non infection control concern
 1. Admitting Dr looked up Fabry's disease on Wikipedia – then we had a 15 minute argument concerning the errors in the wiki article

Clinic visits

- Highly variable hand cleaning in employees
- Highly variable equipment cleaning
 - New neurologist 1st appt & the pin
- Compliance with Airborne/Droplet - a complete gong show (employees & clients)



Hope you liked
our Mom's
presentation

Questions?

Paco

Abby