Case of Lisbeth Bucher

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Question 1

What are the key issues in the dental and medical histories that would likely affect Lisbeth's oral health? What could be the etiology of these issues? What do you need to learn about these issues to inform your dental hygiene care?

Question 1

- Key issues that would likely affect Lisbeth's oral health:
  - Occlusion: Class III profile and obvious anterior open bite with crowding
  - Can only breathe on right side of nose, because the left side is blocked
    - Mouth breather and snores
  - Some type of inherited syndrome
  - Trouble with enunciating
  - Hearing aid in right ear

Question 1

- What could be the etiology of these issues?
  - Differential Diagnoses
    - Pallister Killian Syndrome

Pallister-Killian Syndrome

- General features: profound mental and motor retardation with a seizure disorder, frontotemporal alopecia, rarely diffuse hyper/depigmentation. Slight skeletal defects.
- Craniofacial features: high frontal hairline, low set and dysplastic ears, wide and flat nose bridge, upward slanting palpebral fissures, small nose, with upturned nares, full cheeks, long and simple philtrum with prominent upper lip.
- Oral manifestations: macrostomia with downturned corners, large mandible, high arched palates, and labial pits on lower lip.

Question 1

- What could be the etiology of these issues?
  - Differential Diagnoses
    - Pallister Killian Syndrome
    - Binder's Syndrome

Binder's Syndrome

Characteristic findings: deformities in nasal skeleton, have unusually flat, underdeveloped face, abnormally short and flat nasal bridge, underdeveloped upper jaw with protruding lower jaw or class III malocclusion as well as crowding, hypoplasia of the nose.

- Bilateral loss of hearing
- Mental retardation
- Evidence of family recurrence of the syndrome

Question 1

- What could be the etiology of these issues?
  - Differential Diagnoses
    - Pallister Killian Syndrome
    - Binder's Syndrome
    - Crouzon Syndrome

Crouzon Syndrome

- Intraoral symptoms: anterior open bite, crowding and malocclusion
- Extraoral symptoms: underdevelopment of upper jaw (maxillary hypoplasia)
  - Hearing impairment
  - Possible mental retardation
  - Abnormal deviation of the partition that separates the nostrils (deviated nasal septum) and/or unusual smallness of the air-filled cavities that open into the nose (paranasal sinuses) → upper airway obstruction may result in the need to breathe through the mouth., and abnormalities of bone growth and development → malformations of the craniofacial area, unusually flat or underdeveloped midfacial regions (midface hypoplasia).

Question 1

- What could be the etiology of these issues?
  - Differential Diagnoses
    - Pallister Killian Syndrome
    - Binder's Syndrome
    - Crouzon Syndrome
    - Apert's Syndrome (Working Diagnosis)

Apert Syndrome

- INTRAORAL SYMPTOMS: Class III Malocclusion
  - anterior open bite
  - posterior crossbite
  - supernumerary teeth or missing teeth
  - ectopic eruption (could explain missing teeth)
  - large appearing tongue and a v-shaped maxillary arch
  - Crowding
  - Malocclusion
  - bifid uvula
  - thick gingiva
  - poor oral hygiene
- EXTRAORAL SYMPTOMS: mild mental retardation, brachycephalic skull, sunken middle face, hearing loss

Brachycephalic skull

Question 1

What would we need to learn about these issues to inform your dental hygiene care?

- Need to be informed more about the **'some type of inherited syndrome'** in order to provide the best oral care

    - Consult parents or doctors at Children's Hospital about her condition
    - Communication directly with Lisbeth
    - Look over Lisbeth's past surgeries that helped support her development of bone
    - Life skills program and what it says about her abilities and progress

        - Consent for care

Question 2

What social determinants of health may be influencing the oral and general health of Lisbeth? Develop a list of these determinants of health. Which are the most likely affecting Lisbeth's oral health? Provide the rationale to support your prioritized list

Question 2

Social determinants most likely affecting Lisbeth's oral hygiene:

1. Biology and Genetic Endowment
2. Healthy Child Development
3. Income and Social Status
4. Health Services
5. Education and Literacy
6. Personal Health Practices and Coping Skills
7. Social Support and Network
8. Physical and Social Environment
9. Gender
10. Culture

Question 2

- **Biology and Genetic Endowments**
  - ‘Some type of inherited syndrome’ due to genetics (RF)
  - Differential diagnosis:
    - Pallister-Killian syndrome
    - Binder’s syndrome
    - Crouzon syndrome
  - Working diagnosis:
    - Apert syndrome
      - Mutation of **FGFR2 gene** on chromosome 10q25.3-26
- **Healthy Child Development**
  - Born in/grew up in North America with a middle-class family. Has access to an established healthcare system (DF)
  - Possible poor brushing techniques since she was a child, so those habits followed her through the years (RF)

Question 2

- **Income and Social Status**
  - Both her parents work, provide financial support but unable to attend dental hygiene appointments (DF/RF)
  - Her father will soon be retiring and may be able to come for future appointments but he is hoping to find some other work (DF/RF)
- **Health Services**
  - History of several surgeries on her face to support the development of her bones (DF)
  - The doctors have talked about more surgery and orthodontic treatment, but Lisbeth needs to learn how to look after her mouth better (DF)
- **Education and Literacy**
  - Life skills program at school to help her with communicating, relating to others, setting goals, making and enacting plans, living with and initiating change (DF)

Question 2

- **Personal Health Practices and Coping Skills**
  - Personal health: Apert Syndrome (RF)
  - Self-conscious about the appearance of her teeth (DF)
- **Social Support and Network**
  - Life skills program at school (DF)
  - Grandmother takes care of her (DF/RF)
  - Only child, so lack of a home social-support. Both parents work all the time (RF)
- **Physical and Social Environment**
  - Lives in North America (DF)
- **Gender**
  - Women are more introverted and self-conscious about their appearance (DF/RF)
  - Lisbeth appears somewhat self-conscious (DF)
- **Culture**
  - As a Canadian, values oral and overall health (DF)

Question 3

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How does this new information affect your previous responses?

Question 3

1. Reasons why Lisbeth came to the UBC dental hygiene clinic

- Improve her oral condition before receiving other surgeries or orthodontic treatments.
- Heavy plaque and gingivitis on anteriors (might due to she has anterior crowding).
- Grandmother doesn't have enough oral health literacy.

2. Lisbeth’s entire profile

- Flat/concave profile, generalized acne, underdeveloped midfacial bones, missing teeth, crowding and open bite might greatly affect her confidence.
- Profile characteristics, missing/underdeveloped teeth can help the healthcare professionals indicate what type of inherited syndrome she may have.

Question 4

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What additional information would you like or need to have and how would you go about obtaining this information?

Question 4

1. The Inherited Syndrome

- Contacting Lisbeth's doctors and parents, as well as her parents' doctors.
  - Write a formal letter/email to request information from her previous health care providers.
  - Information being requested could include test information and results (blood, urine etc.); past and future plans for surgeries, family medical history, CT/PET/MRI scans, dental radiographs and other relevant medical information.
  - Ask Lisbeth if she is currently under any kind of medications.

2. Dentition

- Missing Teeth
  - Are they genetically missing, have been extracted or unerupted.
  - Knowing this information by taking radiographs or obtaining information from her previous dentist/ortho specialty and her parents.

3. Dental Hygiene Care

- Her chief concerns.
- Ask her about her oral hygiene routine.

Question 5

When considering the Human Needs Model developed by Darby and Walsh, what unmet needs do you believe Lisbeth may have? What is your preliminary dental hygiene diagnosis based on the information you have? Support each need with possible etiologies.

Question 5

Human Needs Model

- Biologically sound and functional dentition
  - Anterior open bite and unerupted canines can cause difficulties in biting off food with her front teeth, uncomfortable while she is eating
  - Can use her posterior teeth for chewing (no posterior open bite)
- Wholesome facial image
  - Flat or almost concave profile, class III occlusion with prognathic profile
  - Mid-facial bones are not fully developed
  - Generalized acne
  - May feel uncomfortable in front of others or less confident/attractive
- Protection from health risks
  - Concern about safety if currently taking medications from past surgeries or mental condition
  - Assess risk of various infections
  - Check for evidence that she is at risk for oral or systemic disease

Question 5

- Freedom from head and neck pain
  - Some TMJ discomfort due to size and placement of mandible, bilateral enlargement of the alveolar process in the posteriors
  - Tenderness from past surgeries
- Skin and mucous membrane integrity of head and neck
  - Untreated gingivitis especially on anteriors (red and inflamed)
  - Mouth breather- high risk of xerostomia
  - Generalized acne may increase risk of exposure to environmental toxins
- Conceptualization and problem solving
  - Soft spoken and some problems enunciating- some difficulty understanding her speech
  - Uses non-verbal gestures to communicate
  - Unable to determine with certainty how much she understands about oral health
- Responsibility for oral health
  - Brushes teeth two times a day- cares and trying to improve oral health
  - May need parental guidance to aid her in oral self care (but both working)
  - Grandmother does not seem to have high dental IQ

Differential Diagnosis - Binder's Syndrome

- Associated with a deficiency in development in the premaxillary area
- Characteristics include:
  - Deformities of the nasal skeleton and overlying soft tissues
  - Midfacial hypoplasia
  - Abnormally short nose and flat nasal bridge
  - Underdeveloped upper jaw
  - Protruding lower jaw (class III malocclusion)
  - Dental crowding
- Patients can also present bilateral loss of hearing, and mental retardation (no proof)
- Usually isolated but some reports of family recurrence (among siblings or parents and children)
  - 36% positive family history in one study

Differential Diagnosis - Pallister-Killian Syndrome

- Anomaly caused by the tetrasomy of the chromosomal region of 12p
- Characteristics include:
  - Large mandible
  - Macrostomia with downturned corners
  - Low set and dysplastic ears
  - Wide and flat nose bridge
  - Upward slanting palpebral fissures
  - Small nose with upturned nares
- Patients also have profound mental and motor retardation with a seizure disorder, frontotemporal alopecia, and slight skeletal defects

Differential Diagnosis - Crouzon Syndrome

- Rare genetic disorder
- A form of craniosynostosis → fibrous joints between certain bones in the skull prematurely fuse which affects proper development of the skull
- Caused by mutations of one of the fibroblast growth factor receptor (FGFR) genes
  - Normally FGFR2 (which plays an important role in bone growth)
- Symptoms relevant to Lisbeth include:

<u>Intraoral</u>

- Anterior openbite
- Crowding
- Malocclusion

<u>Extraoral</u>

- Maxillary hypoplasia
- Hearing impairment
- Possible mental retardation
- Deviated nasal septum and/or smallness of paranasal sinuses
- Upper away obstruction
- Malformation of craniofacial area → unusually flat or undeveloped midfacial regions (midface hypoplasia)

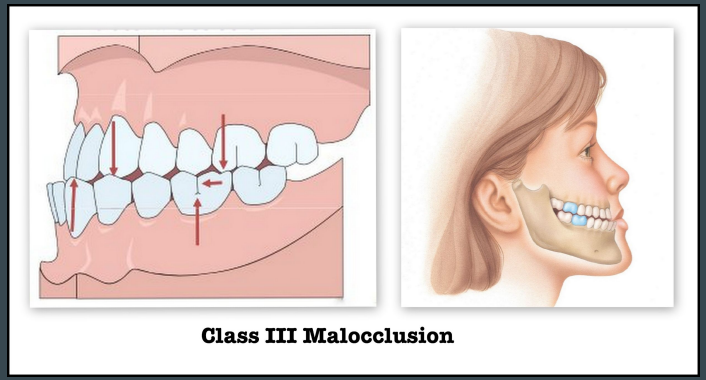
Working Diagnosis - Apert Syndrome

- Rare genetic disorder
  - 1/50,000 babies are born with Apert syndrome
  - 4.5% of all craniosynostosis represent Apert syndrome
  - Can also be sporadic
- Characterized by craniosynostosis, midface hypoplasia, and symmetrical webbing of fingers and toes
- Caused by mutation on FGFR2 gene
  - Plays important role in bone growth and development
  - Mutation rate increases with paternal age
- No cure, but surgery can help correct some of the problems that can occur

Apert Syndrome - Symptoms

Intraoral:

- Class III malocclusion
- Anterior open bite, posterior crossbite
- Supernumerary teeth or missing teeth
  - Lisbeth is missing her first premolars
- Ectopic eruption
  - Lisbeth's radiographs show erupting canines
- Large appearing tongue and v-shaped maxillary arch
- Crowding
- Bifid Uvula
- Thick gingiva
- Poor oral hygiene



Class III Malocclusion

Apert Syndrome - Symptoms

Extraoral

- Long head, high forehead
- Wideset, bulging eyes
- Mental retardation
- Brachycephalic skull
  - Short, broad skull, protruding lower jaw
- Maxillary hypoplasia
- Sunken middle face (midface hypoplasia)
- Hearing loss
  - From many sinus infections
  - Lisbeth wears hearing aid
- Symmetrical webbing of fingers and toes

Question 6

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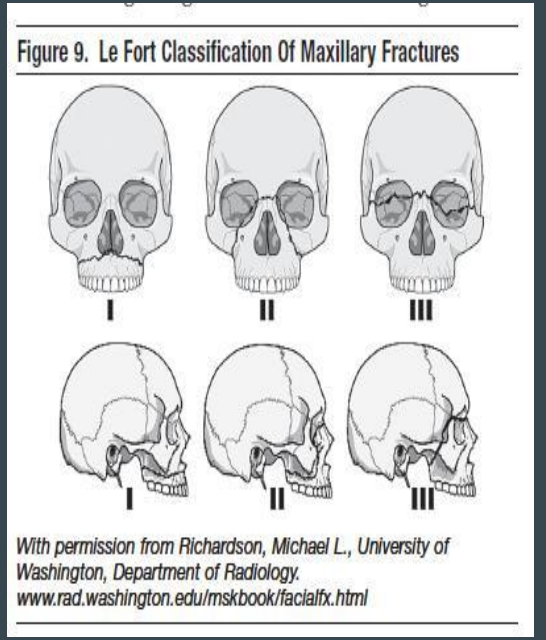
How does this information affect your previous analysis? Is there anything you wish to add or delete from your previous analysis?

Question 6

- Our differential diagnosis included four syndromes
  - ~~Binder's Syndrome~~
  - ~~Pallister-Killian Syndrome~~
  - ~~Crouzon Syndrome~~
  - Apert Syndrome
- From the radiographs we notice:
  - Permanent canines unerupted
    - Primary canine in Quadrant I retained
  - Permanent second premolar partially erupted
  - Central incisor in Quadrant I rotated mesially
  - Third molars in Quadrant II, III, and IV are impacted
  - Tooth in nasal cavity?
  - Crowding is confirmed in maxillary arch

Question 6

- We have confirmation of her past surgical history
  - She is being monitored at the Cleft Palate/Craniofacial clinic for many aspects
    - Plastic surgery, orthopedic, otolaryngology/otology, orthodontics, speech and language
      - Unaware of relation between hands and Apert's
  - Had at least two surgeries with Halos and mid face distraction
- She will be undergoing surgeries in the future
  - LeForte I maxillary advancement to help with class III malocclusion, facial asymmetry, obstructive sleep apnea, maxillary atrophy
    - Grandmother will go in and move her if she thinks she has stopped breathing
  - May get bone graft at right zygomaticomaxillary complex for some residual hypoplasia



Question 6

- More on future surgeries
  - Unrepaired submucous cleft palate
    - Confirmed, uvula bifid but palate elevated well on phonation
    - Maxillary advancement may impact her speech (nasal air emission may increase)
    - The only concern with a bifid uvula is that there are rare occasions when a "submucous" (under the tissue lining) cleft palate occurs, meaning that the midline fusion occurred but did so incompletely. In that situation, if an adenoidectomy (removal of the adenoid) is performed, the palate might not work well in closing off the nose from the mouth during speaking and swallowing
      - Adenoids are glands located in the roof of the mouth, behind the soft palate where the nose connects to the throat.
      - Frequent throat infections can cause the adenoids to enlarge. Enlarged adenoids can obstruct breathing and block the Eustachian tubes, which connect your middle ear to the back of your nose
- Able to understand most of what is in the LA Life Skills program

Question 7

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Would this situation be described as an ethical issue or an ethical dilemma? Why or why not?

Question 7

- The doctor's wife is a part of the email, which breaches confidentiality
  - Lisbeth's personal information and medical history is exposed to a third party

- The situation becomes an ethical dilemma
  - Choice 1: Contact the college of the doctor and inform them about the situation. Breaching confidentiality by showing evidence of the emails about personal/medical information
  - Choice 2: Do nothing

Question 8

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What ethical principles, identified in the CDHA Code of Ethics document and from Darby & Walsh, apply in this situation?

Question 8

Beneficence → want to do what's right for client; useful information in the email

Accountability → could get yourself in trouble for not reporting; must own up to where you received the information

Autonomy → own choice of your actions when provided with the email

Integrity → have to stay true to CDHA's values/morals, as well as your own (it is wrong and breaches confidentiality to send private emails on someone else's account)

Confidentiality → UBC Faculty of Dentistry Confidentiality Statement Patient Information Systems states to report any breaches of confidentiality to system administrator

Question 8

Veracity → to be truthful to yourself as a hygienist with accurate information about the situation and be truthful to the client about their private information

Fidelity → promises to CDHA, loyalty to the patient (and their information), loyalty to the doctor and the acquaintance

Non-maleficence → want to do the least amount of harm to the doctor and patient in order to reach a beneficial outcome

Professionalism → keep a professional attitude towards CDHA, the doctor (and wife), and the patient

Justice → The law and justice system would be practiced with the same rules even though the doctor is in a high status and also an acquaintance

Question 9

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What are the implications of Lisbeth's current oral and general health status for her future status as she ages? What future human needs might she have as she ages?

Question 9

1. Before the surgery and orthodontic treatment:
- Difficulty in communicating with others because of her soft-spoken, muffled voice.
- Difficulty in understanding others and the materials in class because of the mental retardation.
- Less confident in how she looks due to the anterior open bite, flat/concave profile, generalized acne and anterior crowding.
- Poor oral hygiene may develop periodontal disease.

2. During the surgery and orthodontic treatment:
- Missing schools during surgeries.
- Pain and swollen on her face may present during the surgery treatment.
- Braces may cause development of plaque, gingivitis, caries..

3. After the surgery and orthodontic treatment:
- Side effects from post-op medications.
- Missing a lot of school during healing.
- Nasal air emission might be increased, speech problem increased.
- Improved confidence about her looking after the orthodontic treatment and surgeries.
- Mental retardation and the presence of hearing aid might affect her career when she ages, reduced confidence.

Question 9

1. Freedom from Stress and Fear:

- Might feel anxious and scared in the oral healthcare environment due to her slight mental retardation and speech impediments.

2. Wholesome Facial Image:

- After the surgeries there might be scars and a little bit swollen present.
- Young adolescent, due to hormonal effect there might be breakouts.
- Wearing braces during ortho treatment may cause caries, gingivitis etc.
- Post-op medications that might cause xerostomia, which further could cause halitosis.

3. Skin and Mucous Membrane Integrity:

- Ortho work will be done as she ages so it would greatly affect her oral care (gingivitis and plaque indices).
- Possible xerostomia due to post-op medications.

4. Freedom from Head and Neck Pain:

- Post op and orthodontic procedures might cause head and neck pain.

Question 9

5. Conceptualization and Problem Solving:
   - Mental retardation and communication problems will lead to difficulty in understanding of the dental knowledge educated by dental health professionals

6. Responsibility for Oral Health:

   - Difficulty in opening the mouth after surgeries.
   - Soreness from ortho work
   - Low dexterity
   - Difficulty understanding

Question 10

What role can you as a dental hygienist play in Lisbeth’s treatment?

Question 10

- Educator
  - Show proper way of brushing and flossing
  - Recommend appropriate dentifrices and mouth rinses
  - Educate guardians and partake in what can be done at home
- Clinician
  - Clean areas of the tooth that cannot be reached by a toothbrush
  - Come up with treatment plans
  - Set a PSC with goals and guidelines

Question 10

- Researcher
  - Evidence based decision making
    - Research more about Lisbeth's syndrome to have a good understanding and provide a better diagnosis/treatment plan
- Health promoter
  - Promote health lifestyles for longevity
- Advocator (change agent)
  - Collaborate interprofessionally (ortho, doctor, nutritionist, speech therapist, otolaryngologist, plastic surgeon etc)
  - Support clients' rights
  - Advocate for clients by communicating with other healthcare professionals

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