Case history

A 38-year-old woman visits the dentist with her 10-year-old son for his check-up. The dentist tells the mother that her son's gingiva is a bit red. The boy's mother responds that the gingiva often bleed when she brushes her son's teeth, but that she never notices that her son is in pain.

Medical history

The patient has Down syndrome, but there are no specific medical complications. He is not able to brush his teeth and thus, his parents brush his teeth twice a day. He does not like this and sometimes offers resistance.

Dental history

The patient visits the dental practice four times per year. He sees the dentist twice per year for a regular check-up and he visits the dental hygienist twice per year.

Apart from the specific characteristic abnormalities for patients with Down syndrome there are no other complications.

Mouth inspection

In the whole upper jaw and in the molar region of the lower jaw, the gingival is swollen and bleeds on probing. There are no caries.

Questions

1 What causes Down syndrome?

2 What are the possible physical characteristics of a patient with Down syndrome?

3 How is the diagnosis confirmed?

4 Which health problems are associated with the Down syndrome?

5 What is the life expectancy?

6 The patient has gingivitis. What factors enhance the possibility of having gingivitis in patients with Down syndrome?

7 What suggestions can you give to the carer of these patients?

Answers

1 Down syndrome is caused by chromosomal abnormality and can occur in three forms: trisomy 21, translocation and mosaicism (1).

Trisomy 21 has three instead of two chromosomes. In each cell, 47 chromosomes are found instead of the normal 46. The three chromosomes are due to the failure of segregation of a pair of number 21 chromosomes. It is not inherited, but the incidence is correlated with increased maternal age (1). For example, women older than 45 have a one in 35 chance, whilst women younger than 30 years of age have a one in 1000 chance of conceiving a child with Down syndrome (1).

Translocation is hereditary. It is due to the breaking of a piece of chromosome in pair 21 and attaching to other chromosomes. Genetic counselling may help determine the origin of the translocation, but most cases are due to a chance event (1). Approximately 3–4% of the cases of Down syndrome are because of translocation whilst mosaicism occurs only in 1%. Mosaicism is due to an error in one of the first cell divisions, right after conception (2).

2 There are about 50 different physical characteristics observed in children with Down syndrome, but not every child manifests all characteristics.

The most common characteristics are:

- the skull appears small;
- underdeveloped nose with flat nasal bridge;
- eyes present with prominent folds of skin extending from the root of the nose to the median end of the eyebrow which gives eyes an upward slant;
- low muscle tone;
- short stubby fingers with sometimes a single transverse palmar crease;
- mouth and ears are small;
- protruding tongue;
- less hair;
- arms, legs and neck are shorter;
- tendency to be overweight (3, 4).

3 Diagnosis is by checking the chromosomes with a blood test.

4 People who have Down syndrome may have various medical problems of which congenital heart disease is the most common and serious. Of newborn children with Down

syndrome, 40–50% have cardiac problems (5). Multiple cardiac abnormalities exist in around 30% of patients.

- Orthopaedic problems like cervical spine instability and curvature of the spine are often the result of low muscle tone.
- Thyroid problems, mostly hypothyroidism, are seen in 50% of elderly persons with Down syndrome (5). The symptoms are delayed growth, lethargy, obesity, dry skin and short stature (3).
- Hearing loss can be found in 66–89% of more than 15–20 decibels, in at least one ear. The external ear and the bones of the middle and inner ear may develop differently in people with Down syndrome. Hearing loss is generally not severe, but ear infections are common.
- Vision impairment (squinting) can be caused by lack of muscle tone which influences the coordination of the eyes. Cataracts occur (3).
- Immunodeficiency and impaired host defence are frequently seen (3–5).
- Seizure disorders are seen in between 5% and 13% of children with Down syndrome. Anti-epileptic medication is the treatment of choice.
- Leukaemia is more likely to develop (10–15 times more than in a general population) in the early years of life (5).
- Oral manifestations are a protruded tongue due to an underdeveloped maxilla, narrow palate, enlarged tonsils, adenoids and mandibular prognathism; the latter create a small oral cavity. The teeth are generally small and missing teeth and delayed eruption is more frequent in persons with Down syndrome than in the general population (3, 5). Roots are generally shorter and malocclusion is often seen (3). These patients have a high incidence of periodontal disease because of the impaired host defence mechanism and xerostomia. Xerostomia is due to the greater likelihood of developing chronic respiratory infections which will stimulate mouth breathing.

5 Life expectancy depends on the medical problems, especially in the first years of life. After this critical period, the mean age of life is 47 years and over (1).

6 Gingivitis is not only caused by poor dental hygiene, but in the case of a patient with Down syndrome it is also stimulated by mouth breathing and the susceptibility to disease.

7 Naturally good oral hygiene care may prevent gingival and periodontal problems, but carers can also be stimulated to do the following:

- a speech pathologist can be of help in the development of speech and the use and control of the tongue.
- stimulate the child to put the tongue behind the teeth, so that the child can close the mouth;
- stimulate breathing through the nose by closing the mouth when the baby sleeps;
- stimulate the infant to use the tongue by breastfeeding instead of using a bottle.

References

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