Jan Douwes Visser

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Symptoms, Differential Diagnosis, Supplementary Assessment and Treatment

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Preface

In practice, we do not tend to think in terms of differential diagnosis. We often consider the most probable diagnosis based on the patient's complaints. Only when the most probable diagnosis is not supported by physical examination and/or supplementary tests, do we further consider the other possibilities. In this book, *Pediatric Orthopedics: Symptoms, Differential Diagnosis, Supplementary Assessment and Treatment*, we are working towards a system for differential diagnosis based on the principal complaints and findings. In some cases there are more than one different kinds of complaint. As an example, in Osgood-Schlatter, there are complaints of pain and swelling. In those cases we consider the most preveland complaint which in Osgood-Schlatter is pain. In other cases, for instance a meniscal tear, there may be several individual complaints or a combination of these such as pain medially or laterally combined with limited extension and/or swelling. A torn meniscus appears in several differential diagnoses. A system has been established for every diagnosis as described here: (a) explanatory notes, (b) advice on supplementary tests, (c) advice as to which problems can be treated in primary care, (d) when to refer for secondary care treatment, and (e) what the options are for secondary care treatment. Generalized neurological disorders and neuromuscular disorders such as in spasticity, spina bifida, and acute traumatic lesions have been left out. There are many diverse treatment possibilities available. However, many pediatric orthopedic treatments are not evidence based or sometimes have a low or extremely low value from the literature. The author, advisers, and the publisher are not responsible for faults, omissions, or other implications as a result of the information given in this book. Application of the information given in this publication remains the responsibility of the clinician involved.

Groningen, The Netherlands — Jan Douwes Visser

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Abbreviations

ANF	Antinuclear factor
C	Cervical
CRP	C-reactive protein
CT	Computed tomography
ESR	Erythrocyte sedimentation rate
Hb	Hemoglobin
HLA	Human leukocyte antigen
Ht	Hematocrit
L	Lumbar
MRA	Magnetic resonance arthrography
MRI	Magnetic resonance imaging
n.	Nerve
NSAID	Non-steroidal anti-inflammatory drugs
S	Sacral
T	Thoracal
WBC count	White blood cell count

Contents

Chest Wall

1

Chest Wall Deformity

Complaint: there is a deformity of the chest wall.

Assessment: deformity of the chest wall.

Differential diagnosis:
pectus carinatum (pigeon breast)
pectus excavatum (funnel chest)
fusion disorder of the sternum
absent ribs
Poland syndrome

Explanatory note: **pectus carinatum.** The sternum and the adjoining cartilage and bone are anteriorly prominent (Fig. 1.1). The abnormality is most noticeable in the first year of life, but can also develop during puberty (Fig. 1.2). As a rule, pigeon breast is only a cosmetic issue. Respiratory difficulties may occur when the sternum shifts strongly forward, causing the thorax to be in a continuous inspiratory position.

Pectus excavatum There is an indentation on the front side of the chest. The deepest area lies at the level of the distal part of sternum and the xiphoid process (Fig. 1.3). The abnormality is usually present at birth, but can also develop later. A funnel chest seldom causes physical complaints, the issue tends to be only cosmetic. The abnormality is common in Marfan syndrome.

Fusion disorder of the sternum Partial fusion disorders of the cranial area are the most common. In the distal area they are very rare. It is also possible that the sternum is split along the entire length (total sternal fissure). This abnormality is accompanied by cardiac anomalies and/or defects in the diaphragm.

Absent ribs In this abnormality several ribs are usually absent on one side of the chest wall. There may also be sternal and vertebral abnormalities such as hemivertebrae and block vertebrae. As a result there is a flail thorax, which can lead to a shortness of breath.

Poland syndrome In Poland syndrome[1] there is a unilateral absence of the pectoralis minor muscle and the sternal part of the pectoralis major muscle (Fig. 1.4). Males are affected in 70% of cases. It is generally combined with abnormalities of the hand on the same side, such as small hands (hypoplasia) with absent fingers, webbed fingers (syndactyly) and/or shortened fingers (brachydactyly). The combination of hypoplasia of the hand, syndactyly and brachydactyly is sometimes called symbrachydactyly. There may also be absence of the forearm flexor muscles and the entire arm may be under-developed. Associated anomalies may be the Klippel-Feil syndrome[1] (short neck), Möbius syndrome[1] (paralysis of the facial muscles), a Sprengel deformity (elevated shoulder blade) and pectus excavatum.

[1] See Appendix.

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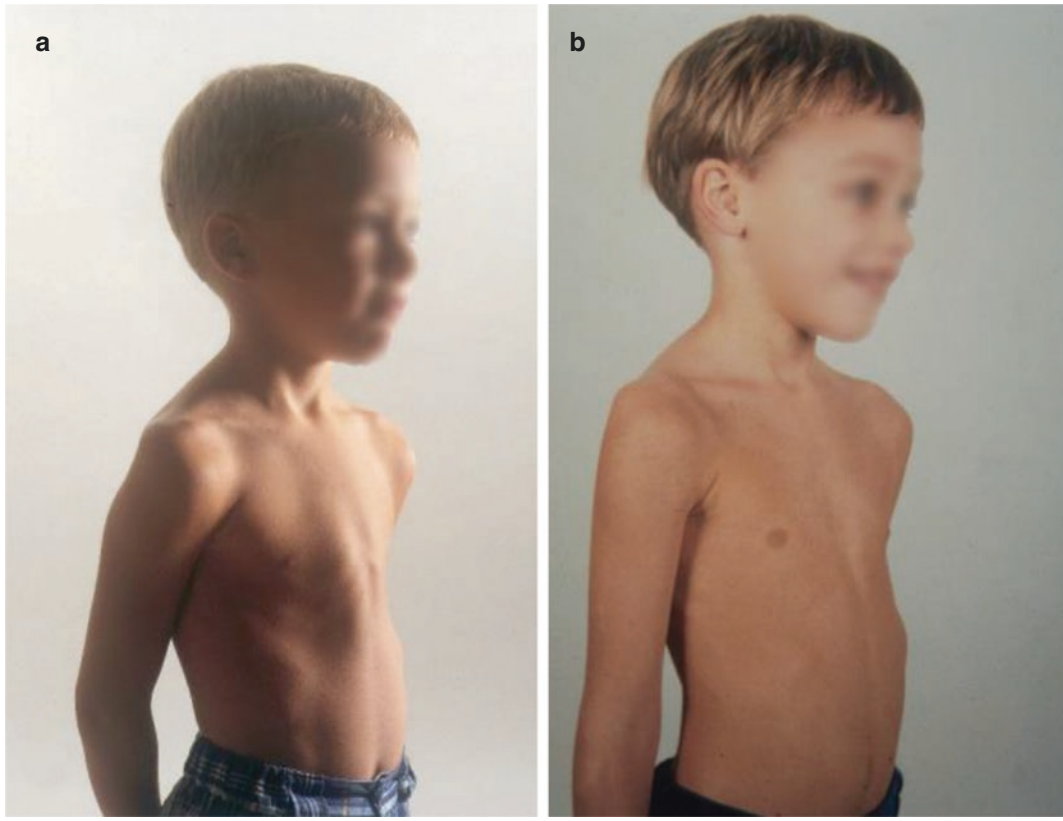

Fig. 1.1 (**a**) Boy, about 5 years old, with a pectus carinatum (pigeon breast). (**b**) After treatment with a trunk orthosis with a pressure pad

Supplementary assessment: in a fusion disorder of the sternum, cardiac abnormalities or defects in the diaphragm should be ruled out.

Primary care treatment: none.

When to refer: the child should be referred as quickly as possible when a shortness of breath accompanies absent ribs. If treatment of the pigeon breast is desired for cosmetic reasons, it is mostly carried out starting at the first year of age. Funnel chest treatment is done starting at the age of 6 years. Infants with a fusion disorder of the sternum should be referred as quickly as possible. Referral is not indicated for the chest deformity in Poland syndrome.

Secondary care treatment: **pectus carinatum.** Spontaneous resolution of the pigeon breast may occur before the first year of age. If the pigeon breast is still present after the first year of age, it can be slowly pushed back with the help of a trunk orthosis with a pressure pad at the level of the prominent thoracic area (Fig. 1.5). A period of 1 year is usually sufficient. The intention is to wear the trunk orthosis day and night, and after correction the trunk orthosis should still be worn for some time at night. This may also be an effective treatment for some cases when the anomaly develops during puberty. The assessing physician must be able to manually press the most prominent portion a little posteriorly. If he cannot do that, there is no point in treating with a trunk orthosis. Treatment with a trunk orthosis in adolescents usually has to be continued until full growth has been achieved. A stiff pectus carinatum may be treated operatively (Fig. 1.6).

Pectus excavatum Starting at the age of 6 or 7 years, a pediatric or thoracic surgeon can carry out a Nuss procedure under thoracoscopic guidance.

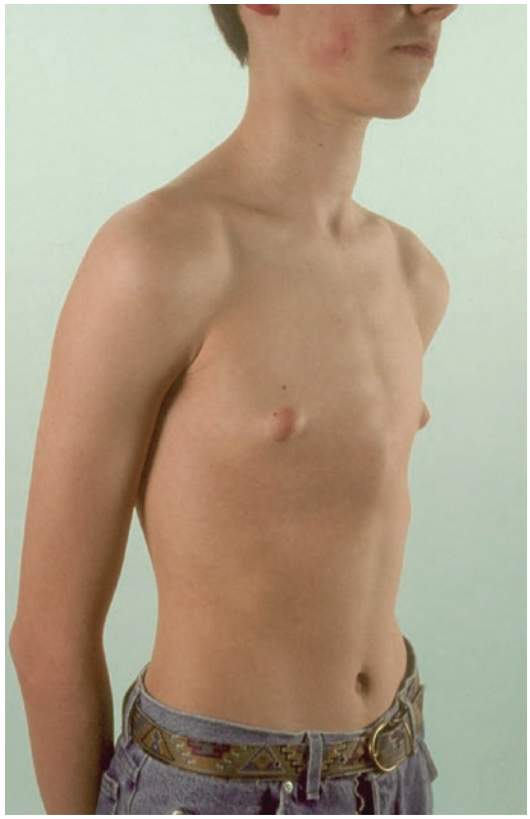

Fig. 1.2 A pectus carinatum (pigeon breast) can also develop during puberty

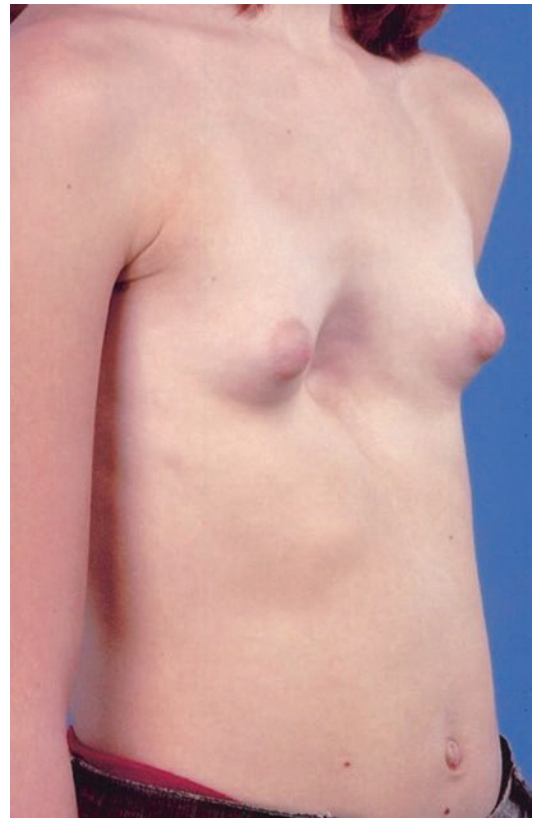

Fig. 1.3 Pectus excavatum (funnel chest)

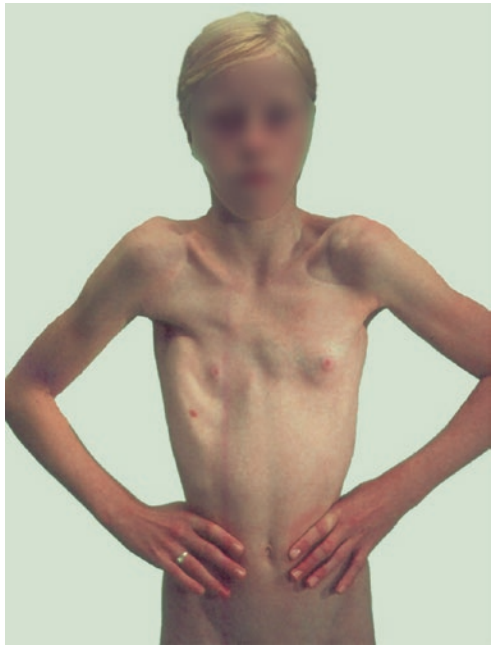

Fig. 1.4 Poland syndrome. On the right side there is a unilateral absence of the pectoralis minor muscle and the sternal portion of the pectoralis major muscle

An incision is made on each side of the chest wall and a forward-curved bar is inserted which acutely pushes the indentation forward (Figs. 1.7 and 1.8). The bar is removed after a few years.

Fusion disorder of the sternum Operative closure of the defect should be carried out as soon as possible after birth.

Absent ribs Early operative correction in the neonatal period gives the best results.

Poland syndrome Treatment of the chest wall abnormality is not indicated.

Chest Wall Pain

Complaint: the child complains of pain in the front side of the chest wall.

Assessment: there is pain on pulpation at the level of the sternum-rib junction.

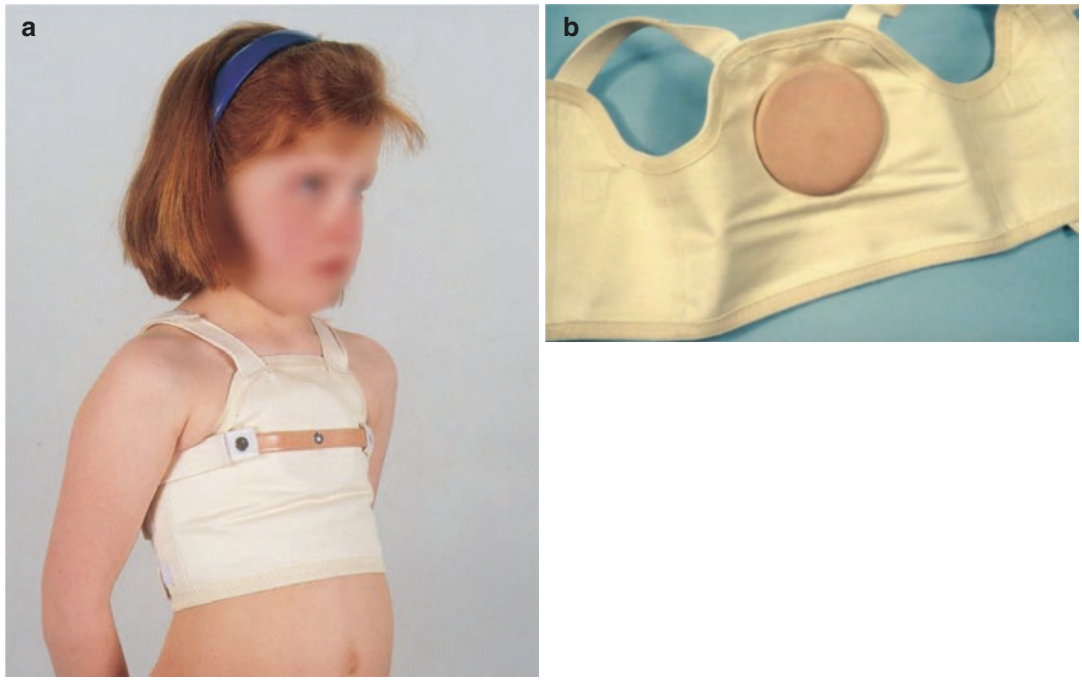

Fig. 1.5 (**a**) Starting at the first year of age, a pectus carinatum (pigeon breast) can be treated using a trunk orthosis with a pressure pad for the prominent thoracic area. (**b**) The pressure pad on the inner side of the trunk orthosis

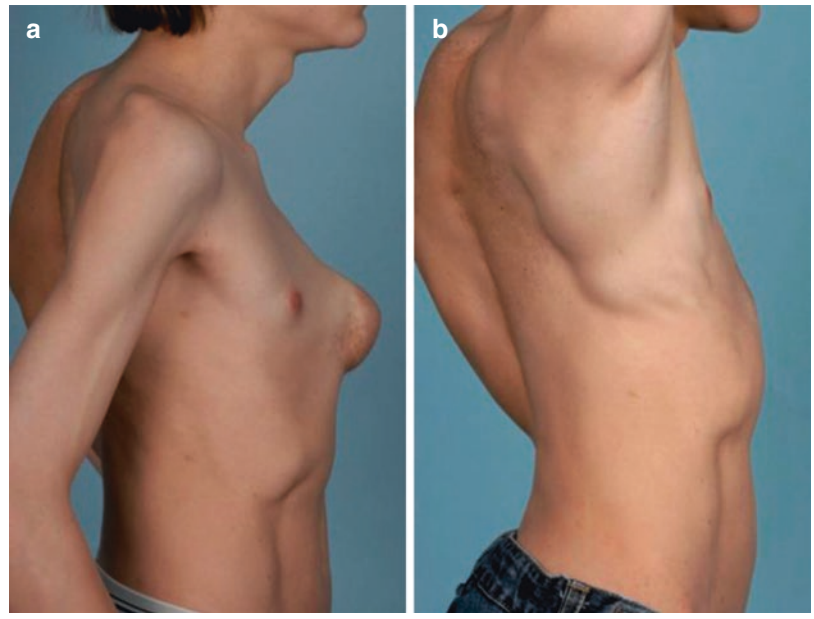

Fig. 1.6 An extensive pectus carinatum (pigeon breast), (**a**) before and (**b**) after operative correction. There is also a considerable scoliosis (Images received from Prof. D.C. Aronson, M.D., Ph.D., The Netherlands)

Differential diagnosis:
Tietze syndrome
costochondritis (costosternal syndrome, costosternal chondrodynia)
bone tumor

Explanatory note: **Tietze syndrome.** In this syndrome, there is a painful unilateral swelling of the rib cartilage at the level of the costosternal junctions. It usually involves the second or third rib. This is a suspected

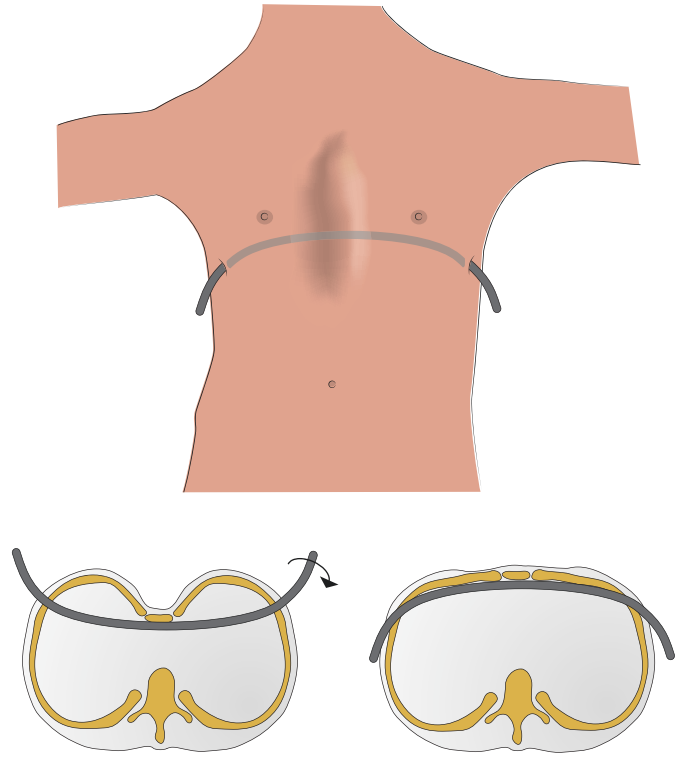

Fig. 1.7 Pectus excavatum (funnel chest): the Nuss operation can be performed starting at the age of 6 or 7. Under thoracoscopic guidance, an incision on each side of the chest wall is made and a curved metal bar is inserted which pushes the indentation forwards. The bar is removed after a few years

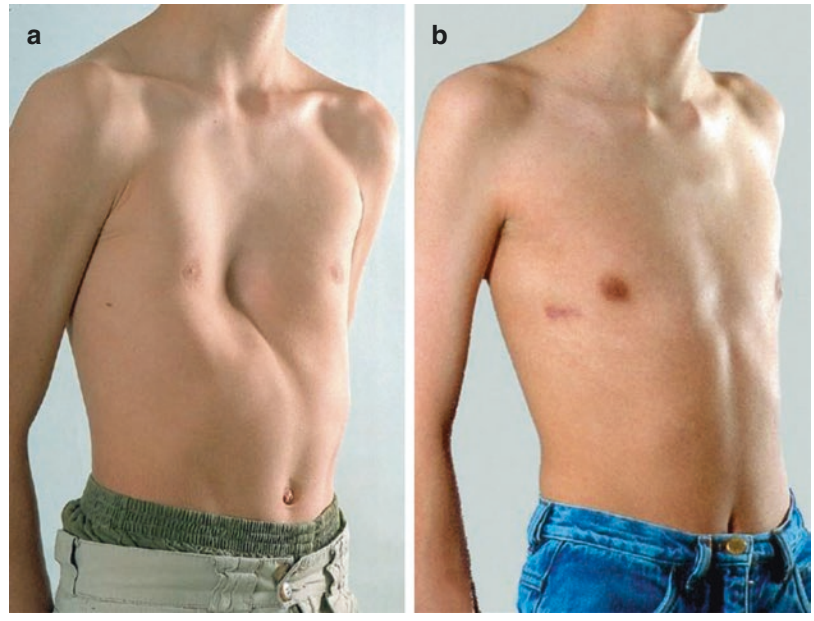

Fig. 1.8 Pectus excavatum (funnel chest), (**a**) preoperatively. (**b**) after the Nuss procedure (Images received from Prof. D.C. Aronson, M.D., Ph.D., The Netherlands)

perichondritis with an unknown cause. Symptoms tend to disappear spontaneously after several weeks to months.

Costochondritis In contrasts to Tietze syndrome several costosternal junctions are involved usually the third, the fourth and the fifth. There is no swelling. The cause is unknown. Pain from costochondritis resolves within a year.

Bone tumor There is painful swelling at the level of the ribs. It is not possible to manualy press the bony swelling posteriorly (see Table 1.1).

Supplementary assessment: If there is any doubt of a tumor X-rays, MRI or CT scan may be requested.

Primary care treatment: In case of Tietze syndrome or costochondritis pain medication may be given if necessary.

When to refer: if a tumor is suspected.

Secondary care treatment: **bone tumor.** A bone tumor should be treated in a medical center specialized in bone tumors. Treatment depends on the nature of the tumor.

Table 1.1 Tumours at the level of the chest cavity. The tumours identified with the § sign are rare

Benign bone tumors	Malignant bone tumors
Osteochondroma	Ewing sarcoma
Enchondroma	
Fibrous dysplasia	
Eosinophilic granuloma	
Aneurysmal bone cyst §	
Osteoid osteoma §	

Based on Adler and Kozlowski (1993)

Differential Diagnosis Chest Wall Deformity

Prominence of the anterior area of the chest wall.	**Pectus carinatum (pigeon breast)**
Indentation of the anterior area of the chest wall.	**Pectus excavatum (funnel chest)**
Split at the level of the sternum.	**Fusion disorder of the sternum**
Flail thorax.	**Absent ribs**
One-sided absence of the pectoralis minor and the sternal part of the pectoralis major muscles.	**Poland syndrome**

Diagnosis: Chest Wall Pain

Chest wall pain	
Painful swelling of the costosternal junction usually the second or third rib.	**Tietze syndrome**
Pain without swelling of the costosternal junctions of the third, fourth and fifth rib.	**Costochondritis**
Painful swelling at the level of the ribs.	**Bone tumor**

Neck

2

Neck Deformity

Neck Flexion Deformity

Complaint: flexion deformity of the neck.

Assessment: instead of a normal cervical lordosis there is a kyphosis.

Diagnosis: **cervical kyphosis**

Explanatory note: **cervical kyphosis.** A cervical kyphosis in a neutral posture should be considered abnormal. Cervical kyphosis can be caused by one or two under- developed vertebral bodies or an operation in which a laminectomy was performed, or it can be part of a syndrome (Table 2.1). If it is one of the symptoms of a syndrome, the child's other abnormalities are usually so impressive that the practitioner has not noticed the kyphosis in the neck. It is however important to identify the cervical kyphosis because even during infancy compression of the spinal cord can already occur, resulting in lifelong paraplegia or even death.

Supplementary assessment: anteroposterior and lateral X-rays as well as a CT-scan and a MRI of the cervical spine.

Primary care treatment: none.

When to refer: all cervical kyphoses should be considered pathological and be referred as quickly as possible.

Secondary care treatment: **cervical kyphosis.** In milder kyphosis without neurological abnormalities a wait-and-see approach may be taken. One should wait until the age of 18 months before performing a spondylodesis for more severe kyphosis without neurological abnormalities.

Neurological abnormalities necessitate earlier intervention. In addition to the cervical spondylodesis, the spinal cord must be decompressed.

Short Neck

Complaint: short neck.

Assessment: the patient has a short neck (Fig. 2.1). In 50 % of cases there is a triad:

- short neck
- limited mobility of the cervical spine
- growth of cranial hair on the back

In less than 20 % of cases extra skin can be seen on the lateral side(s) of the neck with muscular fascial tissue in the form of a wing (pterygium colli), known as "webbed neck". This extra tissue

Table 2.1 Syndromes with a cervical kyphosis. See Appendix for features of syndromes

Campomelic (camptomelic) dysplasia
Conradi-Hünermann syndrome
Larsen syndrome
Neurofibromatosis (Von Recklinghausen disease)

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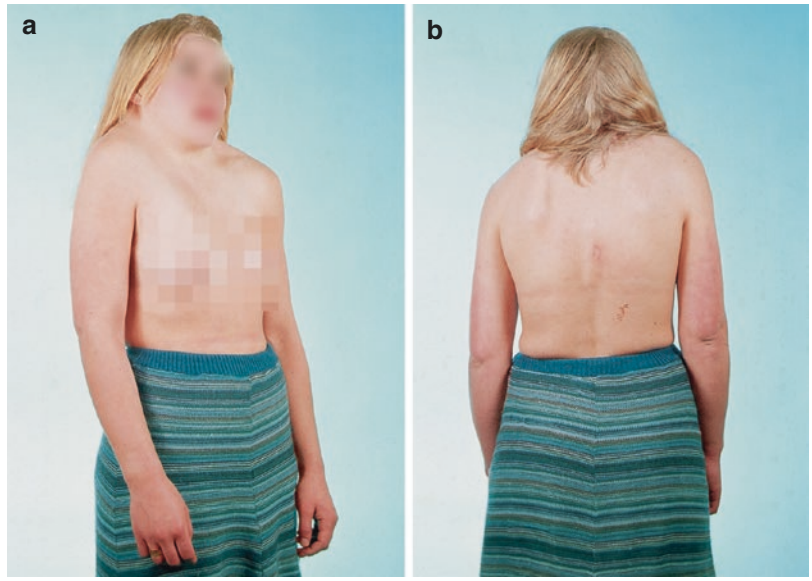

Fig. 2.1 (**a**, **b**) Klippel-Feil syndrome. The short neck and the low cranial hair growth

is spread between the mastoid process and the acromion on both sides.

Diagnosis: **Klippel-Feil syndrome**[1] **(congenital brevicollis)**

Explanatory note: **Klippel-Feil syndrome.** The clinical picture was described by the French physicians Klippel and Feil in 1911 as "l'homme sans cou" (man without a neck). The short neck is caused by vertebrae that have fused together. There may also be a unilateral unsegmented bony connection, hemivertebrae, absence of the posterior elements (arches and spinal processes), a basilar impression, an occipitoatlantal synostosis, an atlanto-axial subluxation and a hypoplasia of the dens. In 20 % of cases the abnormality is accompanied by a torticollis, in 60 % there is a kyphoscoliosis at the thoracic level caused by the hemivertebrae and unilateral and unsegmented bony connections. In 30 % of cases there is an elevated posture and underdevelopment of the shoulder blade, known as a Sprengel deformity. In about 25 % of cases there are cardiac and renal abnormalities, and in 30 % deafness. Neurological complications in adults are possible as a result of compression of the myelum or of the exiting spinal nerves.

Supplementary assessment: X-rays as well as a CT-scan and a MRI of the cervical and thoracic spines.

Primary care treatment: if the patient has no pain or neurological symptoms there will be a wait-and-see policy. If there is pain, it is advisable to limit the activities and a cervical collar can be prescribed.

When to refer: in the early phase to the pediatrician before detecting any other anomalies. Referral should be made to the orthopedic surgeon for additional anomalies such as muscular torticollis and scoliosis, and to the neurosurgeon if neurological complications occur at an older age.

Secondary care treatment: **Klippel-Feil syndrome.** A torticollis based on a shortened sternocleidomastoid muscle can be treated by dividing or lengthening the muscle. Neurological symptoms are caused mostly by an atlantoaxial subluxation, a basilar impression or a congenital occipitoatlantal synostosis (for treatment, see pp. 15, 18, 19).

[1] See Appendix.

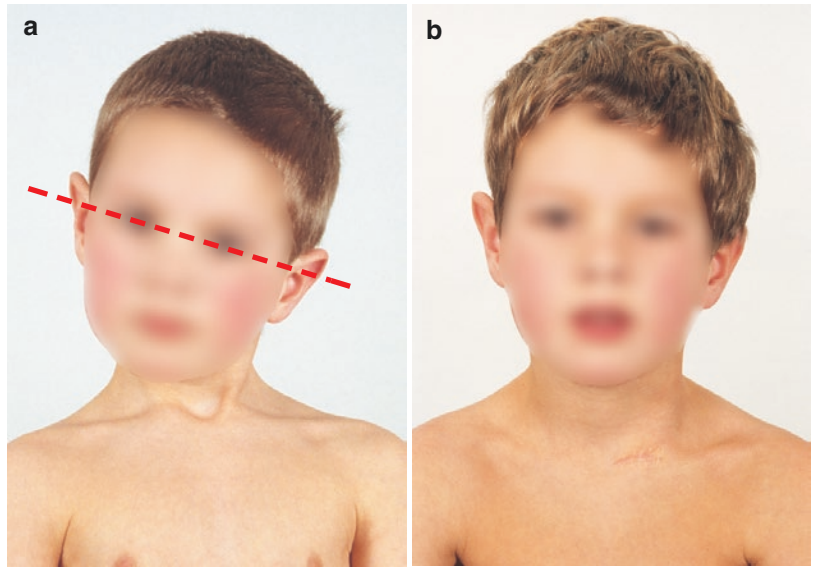

Fig. 2.2 (**a**) Torticollis. Contracture of the left sternocleidomastoideus muscle. An easy way to determine on which side the abnormality lies is to draw an imaginary line between the pupils. The shoulder this line points to is the affected side. (**b**) Situation 2 years after dividing the left sternocleidomastoid muscle origin

Wry Neck

Complaint: a wry neck is usually present at birth, but sometimes it appears at an older age.

Assessment: lateral flexion of the head towards the affected side, the chin is turned towards the non-deviated side. An easy way to determine on which side the deviation lies is to draw an imaginary line between the pupils. The shoulder that this line points to is the affected side (Fig. 2.2).

Differential diagnosis:

torticollis
- infantile muscular torticollis
- (congenital muscular torticollis)
- juvenile muscular torticollis
- osseous torticollis
- ocular torticollis

Explanatory note: **torticollis.** The cause of the infantile as well as juvenile muscular torticollis is unknown.

Infantile muscular torticollis In 20% of cases a swelling in the trajectory of the sternocleidomastoid muscle is visible and palpable between 10 days and 4 weeks after birth (Fig. 2.3). The swelling reduces gradually 4 weeks after it has appeared. By the age of 4–6 months the swelling has disappeared. In 80% of cases the swelling is not observed or not recognized, and the

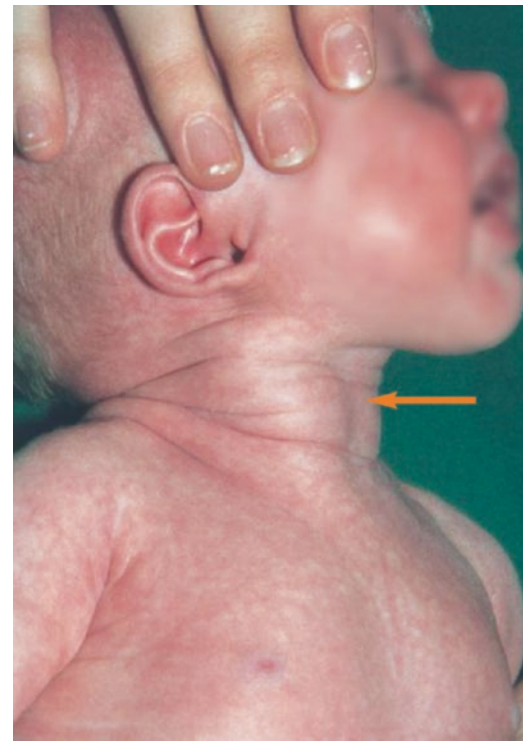

Fig. 2.3 Swelling in the trajectory of the right sternocleidomastoid muscle (*arrow*)

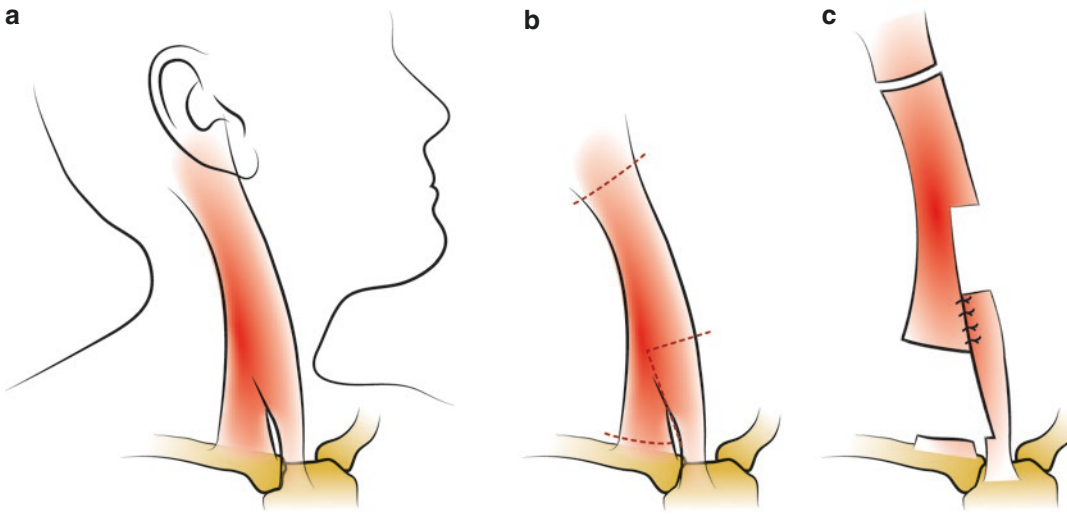

Fig. 2.4 (**a**) The sternocleidomastoid muscle has a sternal and a clavicular origin. The shared muscle belly inserts into the mastoid process and occiput just behind the ear. The sternocleidomastoid muscle can operatively be lengthened or divided at the level of the clavicle and sternum. If there is a recurrence the insertion at the level of the mastoid process and occiput may also have to be divided. (**b**, **c**): In this case the sternocleidomastoid muscle is divided at the mastoid process and occiput. The origins of the clavicular and sternal head are lengthened with a Z-plasty (Redrawn from: Ferkel RD, Westin GW, Dawson EG, Oppenheim WL. Muscular torticollis. A modified surgical approach. J Bone Joint Surg Am. 1983;65-A:894–900)

abnormality is later on identified on the basis of a contracture of one or both heads of the sternocleidomastoid muscle.

The sternocleidomastoid muscle has a sternal and a clavicular origin. The joint muscle belly inserts into the mastoid process and occiput just behind the ear (Fig. 2.4). The cause of the swelling in the sternocleidomastoid muscle is unclear. It used to be assumed that during a difficult birth (breech presentation or forceps delivery) there was bleeding into this muscle, followed by fibrosis of the hematoma. However, blood in the swelling has never been shown. It is now assumed that the fibrous tissue is already present before birth.

Spontaneous recovery of the torticollis occurs during the first year of life in 90 % of cases. If the abnormality persists, the face and the skull will also become deformed, with a flattening of the skull and the face on the side of the contracture (plagiocephaly). A developmental dysplasia of the hip is found in 20 % of children with an infantile muscular torticollis. If there is no contracture of the sternocleidomastoid muscle, the underlying causes are a congenital anomaly of the skeleton or an ocular abnormality.

Juvenile muscular torticollis Sometimes a muscular torticollis appears at childhood. In that case there is a contracture of both heads of the sternocleidomastoid muscle. This condition does not recover spontaneously.

Osseous torticollis Due to the presence of a hemivertebra (half vertebra) or one or several asymmetric nonsegmented bony connections, a wry neck can be present. The patient has Klippel-Feil syndrome[2] if the neck is also too short.

Goldenhar syndrome or oculoauriculovertebral dysplasia is a special form of osseous torticollis in which in addition to abnormal vertebrae there are also eye and ear abnormalities such as epibulbar dermoid cysts and preauricular skin anomalies.

Ocular torticollis This anomaly is present from birth, but is often only noticed around the age of 9 months, after the child has achieved sitting balance. Paresis of the extraocular muscles, generally the superior oblique muscle, causes crossed and double vision when the head is held horizon-

[2]See Appendix.

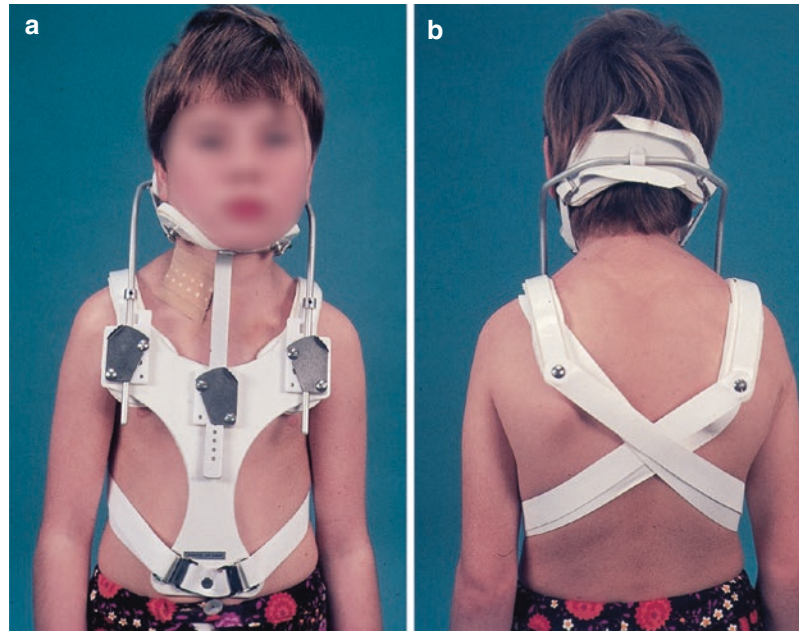

Fig. 2.5 (**a**, **b**) Neck orthosis, in this case a SOMI orthosis (*S* sternum, *O* occiput, *M* mandibula, *I* immobilization)

tally. The child must hold his head slanted in the frontal plane to prevent double vision.

Supplementary assessment: radiological assessment of the cervical spine to reveal or rule out an osseous torticollis. In infantile or congenital muscular torticollis an ultrasound of the hips or an anteroposterior X-ray of the pelvis should be made to check for a developmental dysplasia of the hip.

Primary care treatment: a patient with infantile muscular torticollis is usually referred to a pediatric physiotherapist up to the age of 18 months. The parents do stretching exercises under the supervision of the pediatric physiotherapist. It is not certain whether these stretching exercises influence the natural history. Infantile muscular torticollis disappears spontaneously around the first year of life in 90 % of cases.

When to refer: children with a muscular or osseous torticollis should be referred to an orthopedic surgeon when the child is older than 18 months. If the sternocleido-mastoid muscle is not shortened and there are no osseous abnormalities the child should be referred to an ophthalmologist.

Secondary care treatment: **infantile muscular torticollis.** If the abnormality is still present at 1 year of age there is little chance of spontaneous recovery. In such cases, the sternocleidomastoid muscle should be operatively lengthened or divided at the clavicular and/or sternal origin (Fig. 2.4). The ideal age for operative treatment is in the third year of life. The asymmetry of the face and the skull restores completely after that. Operating before this age increases the chances of an ugly scar and retraction of the skin at the level where the sternocleidomastoid muscle has been lengthened or divided. Even after this age it is not too late to carry out the correction. Good cosmetic results can be achieved up to the age of 12. The sooner the treatment is implemented, the better the ultimate result will be with regard to facial symmetry.

Postoperative treatment involves the use of a neck orthosis day and night for 6 weeks (Fig. 2.5). In the case of a recurrence, in addition to dividing or lengthening the origin at the level of the clavicle and sternum the insertion at the level of the mastoid process should also be divided.

Juvenile muscular torticollis Usually this type is permanent and requires operative treatment.

Osseous torticollis Spondylodesis over a trajectory as short as possible should be carried out.

Ocular torticollis Treatment by an ophthalmologist.

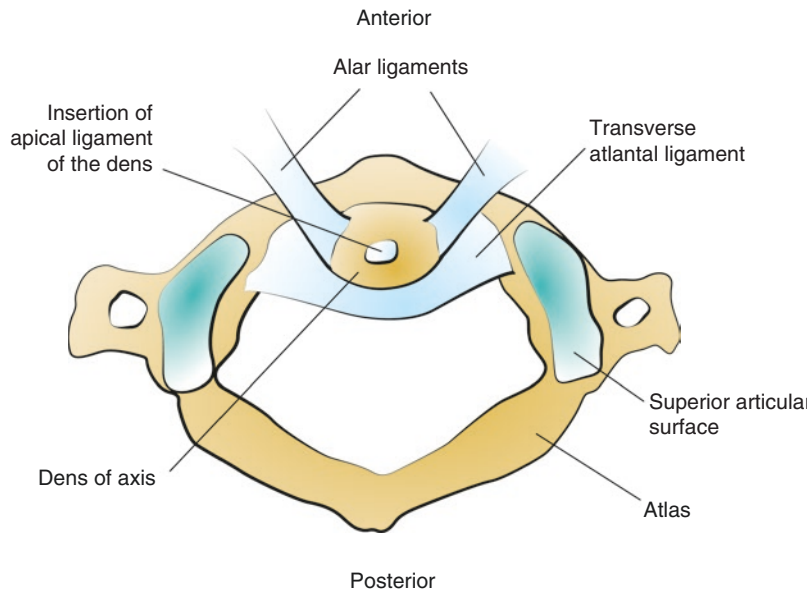

Fig. 2.6 Ligaments at the level of C1 and C2

Neck Pain

Acute Onset of a Painful Stiff and Possibly Wry Neck

Complaint: the child has a painful stiff neck with a possible abnormal posture.

Assessment: there is no mobility in the cervical spine and there may be a wry neck.

Differential diagnosis:

occipitoatlantal subluxation
atlantoaxial subluxation
Grisel syndrome
Sandifer syndrome
spondylodiscitis (discitis)
disc calcification
juvenile idiopathic arthritis

Explanatory note: **occipitoatlantal subluxation.** An occipitoatlantal subluxation is very rare, and is generally caused by abnormal weakness in the ligaments between the dens and the back of the skull and these are the apical ligament of the dens and the alar ligaments (Fig. 2.6). This causes an occipitoatlantal subluxation and occurs in 60 % of cases of Down syndrome[3]. These children are mostly symptom-free.

Atlantoaxial subluxation An anterior atlantoaxial rotatory displacement can arise as a result of a congenital weakness or tear of the transverse atlantal ligament, which is the case in 10–20 % of children with Down syndrome. A posterior atlanto-axial rotatory displacement can occur as a result of an under-developed (hypoplasia) or absent (aplasia) dens (Fig. 2.7). Atlantoaxial subluxations also appear in dwarfism, such as spondyloepiphysial dysplasia[3] (40 %), often accompanied by hypoplasia of the dens, an os terminale or an os odontoideum (Fig. 2.8), and in mucopolysaccharidoses with Morquio syndrome[3] as its most common type and is often accompanied by a hypoplasia of the dens.

Grisel syndrome Grisel syndrome is characterized by the acute occurrence of a stiff and wry neck resulting from a throat infection, such as tonsillitis or a retrotonsillar abscess. The infection weakens the intervertebral ligaments and an atlantoaxial subluxation is the result.

Sandifer syndrome An acute painful stiff and possibly wry neck may occur as a result of

[3]See Appendix.

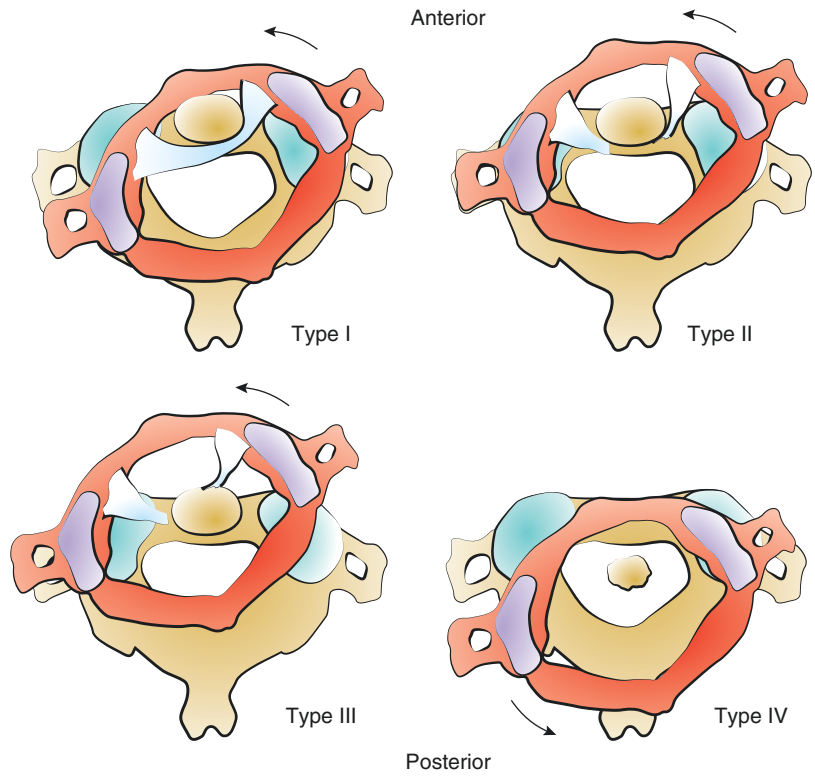

Fig. 2.7 Atlantoaxial subluxation. *Type I*: Rotatory displacement, the transverse atlantal ligament is intact. No anterior shift. *Type II*: Rotatory displacement. One of the two atlantoaxial joints is subluxated. The transverse atlantal ligament is insufficient. Anterior shift of 3–5 mm. *Type III*: Rotatory displacement. Both atlantoaxial joints are subluxated. Insufficient transverse atlantal ligament. Anterior shift of more than 5 mm. *Type IV*: Posterior subluxation. There is hypoplasia or aplasia of the dens (Redrawn from: Fielding JW, Hawkins RJ. Atlantoaxial rotatory fixation. (Fixed rotatory subluxation of the atlanto-axial joint). J Bone Joint Surg Am. 1977;59-A:37–44)

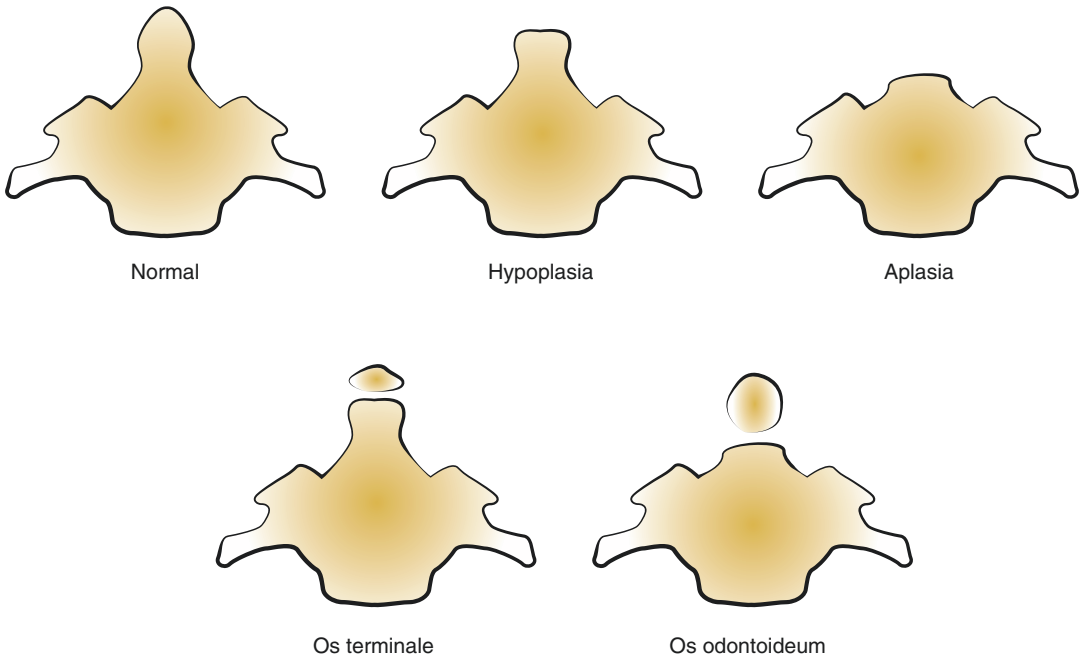

Fig. 2.8 Dens variations

heartburn caused by gastroesophageal reflux in young children, during or right after a meal. There is no atlantoaxial subluxation.

Spondylodiscitis Older children usually have fever in addition to a painful stiff neck. Discitis is generally caused by staphylococcus aureus.