Ocular Anomalies Associated with Syndromes of the Head and Neck

John K. Brooks, DDS

Learning Objectives:

After reading this article, the reader should be able to:

1. Recognize the major criteria of several syndromes of the head and neck
2. Identify some ocular abnormalities associated with the featured syndromes
3. Provide timely referrals of affected patients to appropriate health care personnel
4. Recognize various medical terms of anomalous conditions

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INTRODUCTION

Patients exhibiting dysmorphic features occasionally may present to dental clinicians for examination and treatment. Familiarization with the characteristic facies and of the major criteria requisite for the classification of syndromes may aid with their comprehensive physical assessment and documentation, offer diagnostic utility, improve communication skills with attendant health care practitioners, and perhaps, provide timely referral for therapeutic management (1). A wide range of syndromes manifest abnormalities affecting the eyes (2). Six case presentations of ocular anomalies concomitant with syndromes of the head and neck are illustrated to extend the knowledge to dental professionals.

OCULAR ANOMALIES

Blue sclera is attributed to a primary collagen defect, resulting in scleral thinning and translucency (3). As a consequence, the underlying pigmentation and vascularity of the uvea imparts a blue to blue-gray hue to the sclerae. Blue sclerae are associated with various inherited disorders of the head and neck (Table 1). Prednisone and minocycline administration and iron deficiency have also been reported to produce blue sclera (14-16). Furthermore, the sclerae of healthy newborns may also appear mildly blue, but will usually disappear by the age of 6 months as the scleral tissue thickens.

Figure 1. Gray-blue sclera associated with osteogenesis imperfecta (Case 1).

Figure 2. Hypertelorism, wide nasal bridge, bulbous nose with anteverted nares, thin vermillion border of the upper lip, flat philtrum, and a prominent lower lip seen with the Opitz syndrome (Case 2).

One syndrome linked with blue sclerae is osteogenesis imperfecta, a heterogeneous group of disorders caused by mutations in the genes COL1A1 and COL1A2, which encode type I collagen synthesis (17). Affected individuals may experience an increased frequency of bone fractures, hearing loss, deformities of the extremities and spine, joint hyperextensibility, and increased bruising of the skin. Dentinogenesis imperfecta and opalescent teeth are commonly seen.

Case 1. A 37-year-old female, diagnosed with osteogenesis imperfecta, elicited a history of chronic bone fragility (exceeding 15 fractures). Oral examination was significant for generalized severe caries. Bilateral gray-blue sclerae were evident (Figure 1).

Hypertelorism refers to the abnormal radiologic finding of increased bony interorbital distance. The soft tissue counterpart to this anomaly is termed telecanthus. Although these terms are sometimes used interchangeably in the extant literature, there can be variances between these two measurements. At least 100 syndromes of the head and neck have been identified with increased ocular width.

An example of a syndrome with hypertelorism is the Opitz syndrome, considered to be an autosomal dominant disorder, due to a 22q11.2 deletion, and an X-linked disorder, due to mutation in the M1D1 gene (18, 19). Each variant is associated with multiple congenital midline anomalies and characterized by hypertelorism, laryngeal/tracheal/esophageal defects, hypospadias, and often mental retardation (20). There is an increased incidence of neonatal and childhood death, attributed to the onset of dysphagia and consequent aspiration pneumonia. Oral changes include clefting, ankyloglossia, micrognathia, and a high-arched palate (21).

Case 2. A 12-year-old male, with the Opitz syndrome, manifested unusual facies, comprised of hypertelorism with a wide nasal bridge, bulbous nose with anteverted (upturned) nares, thin vermillion border of the upper lip, flat philtrum, and a prominent lower lip (Figure 2). On profile, low-set ears, with a moderate degree of posterior angulation and micrognathia, were apparent. Surgical repair had been performed for a bifid nose and left cleft lip and palate. Intraoral examination revealed ankyloglossia

and a bifid uvula. The patient's childhood was marked by chronic dysphagia, attributed to a congenital V-shaped notch between the arytenoids, but without a true laryngeal cleft. A urologic assessment revealed the presence of hypospadias.

Another syndrome that features hypertelorism is cleidocranial dysplasia. The mode of inheritance is autosomal dominant and attributed to a defect in the CBFA1 (Core-Binding Factor A) gene, mapped to chromosome 6p21; approximately 20 to 40 percent of cases are attributed to spontaneous mutations (22). Affected individuals may have congenital absence or hypoplasia of the clavicle, either unilaterally or bilaterally. As a consequence, there is an increased range of shoulder motion, allowing the patient to closely approximate the clavicles in front of the chest. Short stature is common. Cranial abnormalities are frontal bossing (prominent forehead), brachycephaly (short skull), delayed suture closure with wormian bones, and hypoplasia of the maxilla and zygoma. Typical facies include reduced midface, broad base of the nose, often with anteverted nares, depressed bridge, and hypertelorism. There may also be alterations of the pelvis, spine, and the extremities. On oral examination, patients appear partially or completely edentulous. Radiographic assessment, however, reveals pervasive failure of tooth eruption, with numerous impacted permanent and supernumerary teeth (23). A high-arched palate may be present.

Case 3. A 34-year-old female, diagnosed with cleidocranial dysplasia, wore maxillary and mandibular partial dentures. The patient was petite and short in height and demonstrated hypertelorism, shortened midface development, and an upturned nose with a depressed nasal bridge. Although the patient lacked generalized joint hyperextensibility, she could easily approximate her shoulders in front of her chest (Figure 3). A panograph was significant for numerous unerupted permanent and supernumerary teeth.

Malformations of the anterior chamber of the eye (goniodysgenesis) involve abnormal development of the cornea, iris, pupil, and lens, as seen with the Rieger anomaly. This rare ocular malformation features hypoplasia of the iris (often leading to pupillary defects), iridocorneal adhesions, and an anterior displaced Schwalbe's line (posterior embryotoxon). The mode of inheritance is autosomal dominant and implicated with mutations with the PITX2 gene along chromosome locus 4q25, the FOXC1 gene along the 6p25 locus, and other genes (24). Additional ocular findings include megacornea, cataract formation, and juvenile-onset glaucoma. When the Rieger anomaly is associated with an atypical pattern of hypodontia and occasionally, redundant periumbilical skin ("outie" belly button), it is referred to as the Rieger syndrome. The

Figure 3. Hypertelorism, shortened midface development, and an upturned nose with a depressed nasal bridge with cleidocranial dysplasia (Case 3). (Reproduced with permission: Brooks JK, Nikitakis NG. Multiple unerupted teeth. Cleidocranial dysplasia. Gen Dent 2008;56(4):393,395-396).

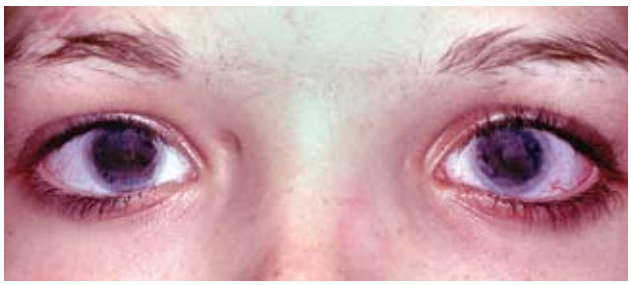

Figure 4. Abnormal pupils and iridal hypoplasia with the Rieger syndrome (Case 4).

pattern of hypodontia with the Rieger syndrome is most atypical and often involves agenesis of the maxillary canines and central incisors. Other dental abnormalities

are misshapen teeth, microdontia, enamel hypoplasia, shortened roots, and delayed eruption (25).

Case 4. A 17-year-old female was diagnosed with the Rieger anomaly, juvenile glaucoma, and a congenital heart murmur. Clinically, each eye exhibited hypoplasia of the iris and slit-shaped pupils (Figure 4). A panographic survey was only remarkable for agenesis of three third molars. The umbilicus was reported to be normal.

Downslanting of the palpebral fissures refers to the clinical finding when the outer canthus of the eye is oriented lower than the inner canthus and featured in at least 30 syndromes of the head and neck. In contrast, the upslanting of the palpebral fissures occurs when the outer canthus of the eye is situated above the inner canthus. At least 17 head and neck syndromes are recognized with this finding and also found as a normal clinical trait with individuals of Asian ancestry.

One such inherited syndrome associated with downslanting of the palpebral fissures is the Marfan syndrome. The mode of inheritance of this connective tissue disorder is autosomal dominant and usually attributed to mutations in the genes that encode fibrillin and TGFBR1 or TGFBR2 on chromosome 15q21 (26). The cardinal features include cardiovascular abnormalities (notably aortic dissection and valvular incompetency), skeletal abnormalities (tall stature with long arms and fingers, laxity of joints), and increased frequency of ectopia lentis (lens dislocation). Diagnosis is paramount as patients are at increased risk for sudden death. Oral manifestations seen are high palatal vault, malocclusion, and micrognathia.

Case 5. A 28-year-old male, with Marfan syndrome, appeared with downward slanting of the palpebral fissures (Figure 5). In childhood, the patient was noted to have pectus excavatum and laxity of the joints. He elicited a history of orthodontic therapy. At present, the patient is tall in stature, with long arms, and displays a high-arched, narrow palate with an underdeveloped mandible (Figure 6).

The Marcus Gunn phenomenon is an involuntary condition where the elevation of a ptotic upper eyelid is prompted by various activities of the mandible (27). This neuromuscular disorder has also been referred to as jaw-winking or pterygoid-levator synkinesis. The term jaw-winking is actually a misnomer as the ptotic eyelid moves in an upward direction rather than downwardly, as in true winking. It has been speculated that neuronal interconnections of the trigeminal and oculomotor nuclei promote simultaneous contraction of the external pterygoid and levator palpebral superioris muscles upon stimulation. The preponderance of cases is congenital in origin, although isolated cases have arisen after trauma and ocular surgical procedures. Mandibular movements that may trigger eyelid elevation include opening of the mouth, chewing, swallowing, sucking, singing, and whistling. In more severe cases, functional and esthetic deficits may be ameliorated with various surgical procedures.

Figure 7. The Marcus Gunn phenomenon (Case 6). Ptosis of the left eye with the patient at rest. (Reproduced with permission: Brooks JK. The Marcus Gunn phenomenon. Discussion and report of a case. Oral Surg Oral Med Oral Pathol. 1987;64(6):687-692.)

Figure 5. Marfan syndrome. Downward slanting of the palpebral fissures (Case 5).

Figure 6. Marfan syndrome. High-arched, narrow palate (Case 5).

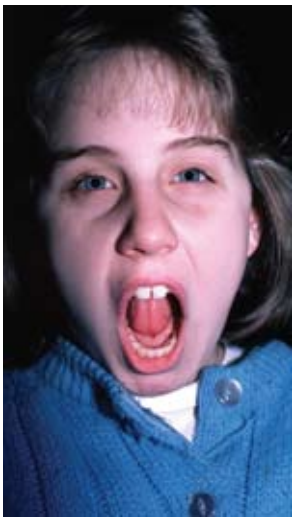

Figure 8. The Marcus Gunn phenomenon. Retraction of the ptotic eyelid upon opening of the mouth (Case 6). (Reproduced with permission: Brooks JK. The Marcus Gunn phenomenon. Discussion and report of a case. Oral Surg Oral Med Oral Pathol. 1987;64(6): 687-692.)

Figure 9. The Marcus Gunn phenomenon. Elevation of the ptotic eyelid with shifting of the mandible to the contralateral side (Case 6). (Reproduced with permission: Brooks JK. The Marcus Gunn phenomenon. Discussion and report of a case. Oral Surg Oral Med Oral Pathol. 1987;64(6): 687-692.)

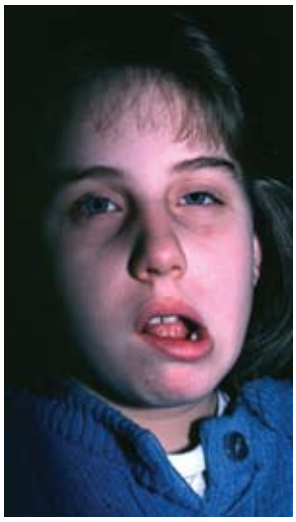

Figure 10. The Marcus Gunn phenomenon. Maintenance of the ptotic eyelid with movement of the mandible to the ipsilateral side (Case 6). (Reproduced with permission: Brooks JK. The Marcus Gunn phenomenon. Discussion and report of a case. Oral Surg Oral Med Oral Pathol. 1987;64(6): 687-692.)

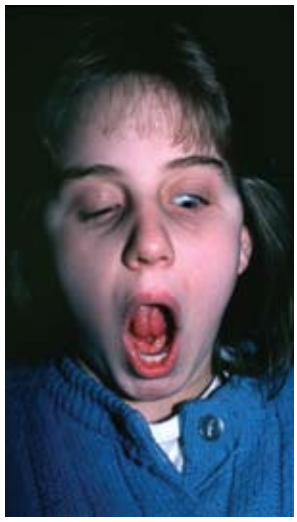

Figure 11. The Marcus Gunn phenomenon. Maximum elevation of the ptotic left eyelid with opening of the mouth with a downward gaze (Case 6). (Reproduced with permission: Brooks JK. The Marcus Gunn phenomenon. Discussion and report of a case. Oral Surg Oral Med Oral Pathol. 1987;64(6): 687-692.)

Case 6. An 11-year-old female demonstrated congenital ptosis of the left eyelid associated with the Marcus Gunn phenomenon. Within days of birth, the patient's mother had recalled that the eyelid would "move more rapidly" as she would suck faster from a nursing bottle. Ptosis of the left eyelid was apparent while the mandible was at rest (Figure 7). Upon opening of the mouth, the left eyelid immediately retracted (Figure 8). Shifting of the mandible to the contralateral side increased the elevation of the ptotic eyelid (Figure 9). However, movement of the mandible to the ipsilateral side maintained the ptosis (Figure 10). Maximum elevation of the ptotic left eyelid was achieved with opening of the mouth with a downward gaze (Figure 11).

CONCLUSION

Six syndromes of the head and neck, accompanied with ocular abnormalities, are presented. While performing a physical examination of the dental patient, attendant personnel should be vigilant for pathologic features of the eyes. Identification of aberrant ocular findings and referral to appropriate health care professionals may offer improved clinical outcomes. Specifically, patients with ptosis may have occult systemic disease, such as myasthenia gravis, hyperthyroidism, progressive muscular dystrophy, and central neoplasms. Moreover, atypical forms of hypodontia with unusual ocular malformations and possibly an "outie" umbilicus may herald the presence of the Rieger syndrome; prompt referral to an ophthalmologist for ocular pressure assessment may avert optic nerve damage from untreated glaucoma. Patients suspected with Marfan syndrome warrant a cardiovascular workup. Previously undiagnosed cases of osteogenesis imperfecta have led to erroneous accusations of child abuse (28). Thus, knowledge of various features in syndromes of the head and neck may aid dental clinicians with timely therapeutic intervention.

SYNDROME	SOMATIC FEATURES	ORAL ANOMALIES
Osteogenesis imperfecta (4)	brittle bones, hearing loss, increased bruising, deformities of the extremities and spine, joint laxity	opalescent teeth, dentinogenesis imperfecta
Ehlers-Danlos syndrome (5)	increased elasticity and fragility of skin, increased bruising, “cigarette paper” scarring, joint laxity, mitral valve prolapse	fragile oral mucosa, Gorlin sign, TMJ subluxation, early onset periodontitis
Marfan syndrome (6)	increased height, elongated torso, lens subluxation, scoliosis, joint laxity, aortic enlargement, mitral valve prolapse, sternal defects, arachnodactyly	high-arched palate, malocclusion, TMJ subluxation
Incontinentia pigmenti (7)	cutaneous lesions, fragile nails, alopecia, mental retardation, seizures	peg-shaped anterior teeth, hypodontia, impacted teeth
Silver-Russell syndrome (8)	facies, short stature, body asymmetry, incurved fifth fingers, café-au-lait spots	downturned corners of the mouth, high-arched palate, underdeveloped jaws
Pseudoxanthoma elasticum (9)	thickened skin with yellowish papules, weakened peripheral pulses, gastrointestinal hemorrhages, mitral valve prolapse	yellow mucosal nodules
Pycnodysostosis (10)	short stature, osteopetrosis, cranial anomalies, shortened terminal phalanges	underdeveloped mandible
Hallermann-Streiff syndrome (11)	facies, hypotrichosis, skin atrophy, short stature, congenital cataracts	micrognathia, natal teeth, hypodontia, supernumeraries, retention of deciduous teeth, severe caries, malocclusion
Hypophosphatasia (12)	deformed and shortened long bones	delayed eruption, premature loss of teeth, alveolar bone loss, enlarged pulp chambers and root canals
Turner syndrome (13)	gonadal dysgenesis, webbed neck, cubitus valgus, short stature	high-arched palate, micrognathia, premature eruption

Table. Syndromes of the head and neck associated with blue sclera. (4-13)

GLOSSARY

arachnodactyly: abnormally long, thin fingers

canthus: corner of the eye

cubitus valgus: deviation of the extended forearm away from the body

Gorlin sign: ability to touch the tip of the nose with the tongue

hypospadias: anomaly of the penis characterized by the presence of the urethral opening on the underside

hypotrichosis: sparseness of hair

iridal: affecting the iris of the eye

palpebral fissure: the space between the borders of the upper and lower eyelids

pectus excavatum: depression of the sternum

ptosis: drooping of the eyelid

wormian bones: small, irregular plates of bone along cranial sutures

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1. **Blue sclerae may result from:**
   A. scleral thickening
   B. scleral thinning and translucency
   C. scleral scarring
   D. lens dislocation

2. **Cardinal features of osteogenesis imperfecta are:**
   A. blue sclerae, brittle nails, dentinogenesis imperfecta
   B. blue sclerae, brittle bones, taurodontia
   C. blue sclerae, brittle bones, dentinogenesis imperfecta
   D. blue sclerae, brittle bones, hypodontia

3. **Medications that may produce blue sclerae are:**
   A. iron, prednisone, and minocycline
   B. iron and prednisone
   C. tetracycline and prednisone
   D. minocycline and prednisone

4. **Anomalies with Opitz syndrome include:**
   A. laryngeal/tracheal/esophageal defects
   B. hypertelorism
   C. hypospadias
   D. all of the above

5. **The Opitz syndrome is considered a:**
   A. multiple congenital midline anomaly syndrome
   B. branchial arch disorder
   C. CHARGE association
   D. pterygoid-levator synkinesis

6. **Oral features of Opitz syndrome include:**
   A. clefting
   B. prognathism
   C. low-arched palate
   D. multiple unerupted teeth

7. **Typical facies with cleidocranial dysplasia are:**
   A. hypertelorism, reduced midface, low-set ears
   B. hypotelorism, reduced midface, anteverted nares
   C. hypertelorism, reduced midface, anteverted nares
   D. hypertelorism, thin pointed nose, microstomia

8. **Patients with cleidocranial dysplasia may:**
   A. be able to approximate their shoulders behind the back
   B. be able to approximate their shoulders in front of the chest
   C. be able to touch the tip of the nose with their tongue
   D. be prone to develop aspiration pneumonia

9. **The Rieger syndrome is often associated with:**
   A. cardiovascular anomalies
   B. tall stature with long, thin fingers
   C. laxity of joints
   D. none of the above

10. **Patients with Rieger syndrome may be at increased risk for:**
    A. juvenile diabetes
    B. juvenile glaucoma
    C. seizures
    D. mental retardation

11. **Common features of Rieger syndrome include:**
    A. ocular anomalies, atypical hypodontia, “outie” belly button
    B. ocular anomalies, atypical hypodontia, “innie” belly button
    C. ocular anomalies, atypical hypodontia, laxity of joints
    D. ocular anomalies, multiple impacted teeth, laxity of joints

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12. The Rieger syndrome may be associated with agenesis of:
A. maxillary lateral incisors and third molars
B. mandibular third molars
C. maxillary central incisors and canines
D. mandibular central incisors and canines

13. Patients with Marfan syndrome may be at increased risk for:
A. aortic dissection
B. lens dislocation
C. TMJ subluxation
D. all of the above

14. The palpebral fissure is:
A. the space between the borders of the upper and lower eyelids
B. the radiographic distance between the orbits
C. the soft tissue distance between the orbits
D. the distance between the inner and outer canthus

15. The Marcus Gunn phenomenon consists of involuntary:
A. closure of a ptotic upper eyelid with opening of the mouth
B. elevation of a ptotic upper eyelid with opening of the mouth
C. elevation of a ptotic upper eyelid with ipsilateral movement of the mandible
D. opening of the mouth with elevation of a ptotic eyelid

16. The Marcus Gunn phenomenon involves miswiring of the:
A. optic and facial nerves
B. optic and trigeminal nerves
C. oculomotor and trigeminal nerves
D. oculomotor and facial nerves

17. Ptosis may be associated with:
A. myasthenia gravis
B. hyperthyroidism
C. central neoplasms
D. all of the above

18. Accusations of child abuse have occurred with undiagnosed cases of:
A. osteogenesis imperfecta
B. amelogenesis imperfecta
C. dentinogenesis imperfecta
D. osteonecrosis

19. Patients with Ehlers-Danlos syndrome often have:
A. increased bruising of the skin
B. decreased elasticity of the skin
C. joint hypomobility
D. Nikolsky's sign

20. Knowledge of features of syndromes of the head and neck may:
A. offer diagnostic utility
B. aid with comprehensive physical assessment and documentation
C. provide timely referral for therapeutic intervention
D. all of the above

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OCULAR ANOMALIES ASSOCIATED WITH SYNDROMES OF THE HEAD AND NECK

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The Dental School is committed to the lifelong education of oral health professionals in Maryland and neighboring states of the Mid-Atlantic region. Continuing Dental Education for dentists and dental hygienists is among the missions of the Dental School; for today's many and frequent advances in science and technology impose a greater and sustaining need for timely accession of new information.

To fulfill its commitment to lifelong education, the Dental School provides courses designed to meet the needs of all oral health care providers. Based on research in the basic and clinical sciences, the Continuing Dental Education Program offers participants educational courses that reflect contemporary professional knowledge of direct benefit to the practice community.

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