Cataract Forms and Grading

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Cataract Types

Partial or complete opacification of the lens is called cataract and can be diagnosed at the slit lamp (Fig. 1). Cataract can be congenital (approx. 1%) or acquired (approx. 99%). Age-related (senile) cataracts are the most frequent cause of acquired lens opacities.

For a better understanding of lens opacities, the anatomical structure and physiological processes are important [1]. The crystalline lens is enclosed in a lens capsule, which is affixed to the ciliary body by the zonula fibres. Once embryonic development completes, this remains a closed space. The anterior lens capsule, which is about 15 µm thick, is lined by a single-layered cubic lens epithelium. The lens epithelial cells divide in the area of the lens equator, where they gradually migrate posteriorly along the posterior lens capsule, which is about 5 µm thick, transforming into elongated lens fibres, and gradually losing the cellular nucleus. These lens fibres form the majority of the lens substance. New fibre cells are deposited in concentric layers, so that the oldest lens fibres are found in the lens core and the youngest in the cortex. This unopposed growth continues throughout life and the lens becomes increasingly dense as the lens fibres in the nucleus increase and the lens becomes increasingly cloudy (Fig. 2).

The lens has multiple functions; it forms part of the dioptric apparatus (about 15–20 diopters), provides accommodation for near vision, and filters ultraviolet radiation (280–400 nm). The lens also separates the anterior and posterior segments of the eye.

All diseases of the lens lead to a more or less pronounced cataract. Risk factors for lens clouding include age, gender, family history, photooxidative stress caused by UV radiation (sunlight exposure), metabolic disorders, nutritional conditions,

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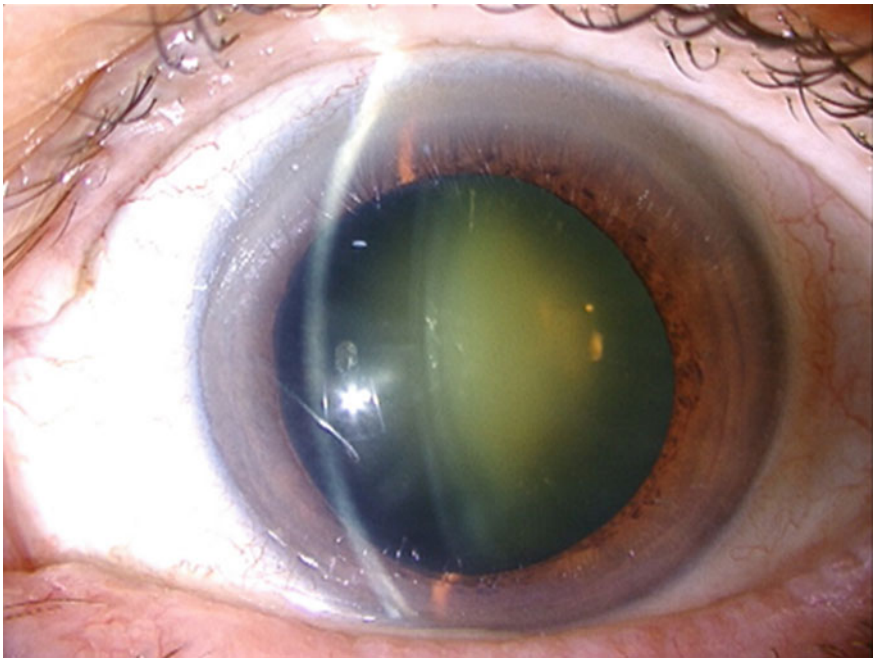

Fig. 1 Nuclear sclerosis in Cat. nuclearis. This form of cataract is very common and can lead to myopia by increasing the central refractive power

alcohol consumption, diabetes mellitus, deficiency of essential amino acids, medications and increased dehydration [1, 2].

The typical symptoms of cataract are a gradual decrease in visual acuity, glare, decrease in contrast vision and monocular double vision [3]. Colour perception may be reduced due to the deposition of chromophores causing a filter effect.

Cataract forms can be classified according to different systems:

(A) temporal occurrence/age (congenital, postnatal, juvenile, senile)
(B) anatomical (nuclear, cortical, subcapsular)
(C) extent (beginning, mature)
(D) etiological (see Table 1 for causes).

(A) Classification of cataract according to age of onset

- congenital—at birth
- postnatal—in the first years of life
- juvenileuntil the age of 30
- senile—all forms associated with increasing age.

In congenital cataracts, heredity and embryonic developmental disorders play the most important role (accounting for approx. 30%) and are usually diagnosed by

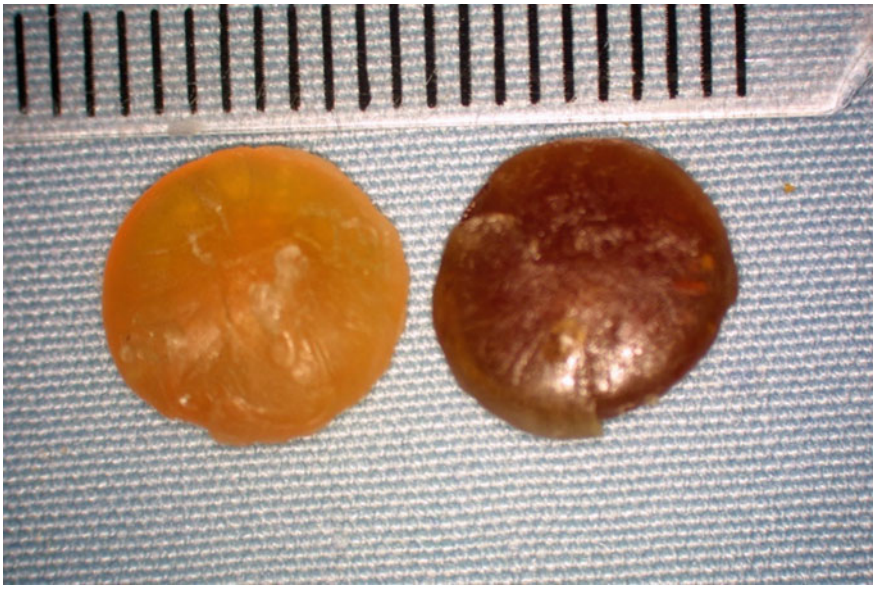

Fig. 2 Brownish discolouration of the lens with increasing clouding. The strong compression in the lens core leads to an increase in the hardness of the lens. These lenses were taken during manual ECCE (left: yellowish-brownish, right: brownish-reddish clouding of the lens)

detecting a leukocoria at screening examinations. Chromosomal causes, as in trisomy 21, and metabolic causes should also be considered (see Table 1). In galactosemia, vacuolisation and swelling of the subcapsular lens fibres occur in the first months of life and are reversible by a diet low in galactose. Furthermore, intrauterine infections, especially in the first 3 months of pregnancy in cases of rubella or toxoplasmosis, are also significant causes of cataract. Rubella infection used to cause cataract more frequently, before the widespread protection by vaccination, as an infection in the first 4 weeks of pregnancy can lead to unilateral or bilateral cataract in up to 50% of cases.

Examples of a congenital cataract are the cataract zonularis/pulverulenta where the clouding of the lens is particularly prominent in the embryonic nucleus (Fig. 3). This risk of severe visual impairment is high due to the dominant heredity and risk of amblyopia (particularly if the cataract is unilateral). Typically, visual acuity improves if the pupil is wide. The more frequent anterior polar cataract is characterized by a central, focal opacity due to a degeneration of the pupillary membrane. A rarer form is the posterior polar cataract (autosomal dominant) with a severe opacity on the rear surface of the lens caused. This is caused by an incomplete regression of the embryonic vitreous vessels (Vasa hyaloidea) or the persistent hyperplastic primary vitreous (PHPV) [1]. Since in many cases the posterior capsule is also defective, there is an increased risk of capsule rupture during cataract surgery in these patients.

Table 1 Rare causes of cataract development

Causes	Type	Features
General	Tetanus	Due to calcium deficiency in hypoparathyroidism occurrence of subcapsular point clouding and radial streaks of the (layered cataract)
	Myotonic dystrophy (Curschmann-Steinert)	Autosomal dominant, dot-like, coloured and white turbidities in the middle cortex, radial spokes, crystallization, rosette figure of the posterior cortex
	Skin diseases (Cat. syndermatotica)	Posterior shell opacity as capsule epithelial cataract in chronic neurodermatitis, more rarely in scleroderma
Metabolism	Diabetes	In type 1 diabetics, there is posterior subcapsular opacity due to metabolic disorders, myopic refraction changes in hyperglycaemia is possible, osmotic changes due to accumulation of sorbitol
	Galactosemia	Galactokinase deficiency, deep posterior cortical opacity due to accumulation of galactite, regression through galactose-free diet
	Wilson's disease	Autosomal recessive, hepatolenticular degeneration in copper storage disease, sunflower cataract due to granular copper deposits in the central lens capsule
Medications	Cortisone	Posterior subcapsular clouding, cataractogenic >10 mg/day for 1 year
Injuries	Siderosis	Brownish opacities due to rust formation (oxidation) of a ferrous intraocular foreign body
	Chalcosis	Sunflower cataract due to greenish intracapsular copper deposits
	Acid-alkaline burns	Different forms of opacities until the lens becomes completely cloudy
Environmental factors	Infrared light	Thermal damage, glass-blowing cataract with fire lamella (true exfoliation on the anterior lens capsule)
	X-rays	Critical dose >2–6 Gy with damage to the epithelium at the equator, clouding of the cortex near the posterior pole after 1–2 years
	Electricity	Vacuoles, axial opacification, anterior, subcapsular

(continued)

Table 1 (continued)

Causes	Type	Features
Syndromes	Down syndrome (trisomy 21)	Cataracta coerulea with flaky cloudiness at the cortex in the equatorial region leads to blue-green colour iridescence
	Patau syndrome (trisomy 13)	Severe ocular malformations with complete opacity of the lens, microphthalmia and coloboma of the uvea
	Alport syndrome	Autosomal dominant, spherophakia due to missing zonula fibres, anterior lenticonus
	Lowe syndrome	X-chromosomal recessive, mikrophakia with small platelike, flattened lens without differentiation of cortex and nucleus (1 mm thickness, 5 mm diameter)
	Fabry disease	Alpha-galactosidase defect, accumulation of glycosphingolipids, Cornea verticillata
Congenital	Coloboma	Indentation at the lens equator inferonasally, missing zonula fibres in this area, usually combined with other coloboma forms of the eye (iris, choroid)
	Spherophakia	
	Microphakia	Lens diameter is reduced and lens thickness increased (spherical lens), refractive myopia, luxation into the pupil with secondary angle block glaucoma (see Weill-Marchesani syndrome)
	Lenticonus/ lentiglobus	Bulging of the anterior or posterior lens pole

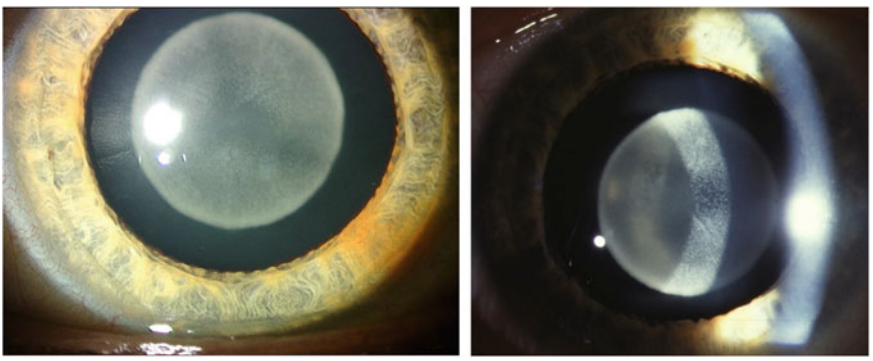

Fig. 3 Cataracta pulverulenta (zonularis) with fine dusty and dot-like opacities and the danger of amblyopia

The most frequent lens opacity in acquired cataracts is the age-related cataract (Cataracta senilis). The development of this form of cataract is multifactorial and can be observed in 5% of patients at 70 years of age increasing to 10% at an age of 80 years. Clouding of the lens in associated with systemic conditions occurs particularly in diabetes mellitus and neurodermatitis. Due to the common embryonic origin of skin and lens from the surface ectoderm, the lens is involved in many dermatological diseases. Iritis/uveitis also leads to a cataract more frequently. Drug-induced cataract occurs mainly during high-dose topical or systemic cortisone therapy. Finally, ocular trauma can also cause cataract.

Note

All diseases of the lens lead to cataract.

(B) Anatomical classification of the cataract

The type of opacity can be assessed using the slit lamp:

- Nuclear opacity (nuclearis) is caused by an increase in the number of lens fibres within the lens, resulting in a significant increase in protein content of up to 30% (Fig. 1). The nuclear opacity leads to a myopic and near vision may again be possible without reading glasses "second sight of the elderly". However, this can be accompanied by monocular double vision and visual acuity may improve at night due to mydriasis.
- Cortical opacity (corticalis) occurs due to wedge-shaped opacities along the lens fibres caused by constant movement during accommodation. Progression is slow in these cases and the symptoms include blurred vision and monocular diplopia. —In posterior subcapsular opacity (subcapsularis posterior), fibrous metaplasia of the lens cells occurs as a result of disturbance in the transformation of the lens fibres from the equatorial zone, i.e. due to metabolic disorders. Rapid progression can occur in these cases and causes severe glare. Near vision in these cases is typically worse than far vision. This form of cataract is common with general diseases, such as diabetes, neurodermatitis and with cortisone therapy.
- Cataracta coronaria (wreath-shaped cataract) with cortical clouding of the cortex
- Cataract coerulea (Blue-dot cataract) with wreath-shaped aquamarine turbidities
- Christmas tree cataract is characterised by colourful, iridescent crystalline clouding due to the inclusion of cholesterol crystals.

(C) Developmental stages of lens opacities

- Cat. incipiens: beginning opacity individual layers.
- Cat. provecta: advanced lens opacity with the indication for cataract surgery
- Cat. intumescens: rapid clouding due to fluid absorption and volume increase, especially in the cortex area with the risk of phacolytic glaucoma if lens proteins escape the capsule.
- Cat. immatura: combination of different anatomical lens opacities (see above).

– Cat. matura: complete opacity of the lens. The lens may be whitish (nivea), brownish (brunescens), reddish (rubra), or black (nigra) in colour (Fig. 2). This classification makes it possible to estimate the hardness of the lens, which can make it difficult to break up the nucleus with phacoemulsification. In cataracta nivea there is often increased intracapsular pressure due to an incipient liquefaction of the lens material with the risk of spontaneous tearing of the entire lens capsule or during capsulorhexis (so-called "Argentinian flag sign").
– Cat. hypermatura (Cat. Morgagni): the lens material is partially liquefied, and the lens core has sunken into the capsular bag, with the risk of phacolytic glaucoma due to tearing of the lens capsule.

(D) Etiological classification of cataract

Although there are many causes for lens opacities, they are particularly common in metabolic disorders (see Table 1).

Cataracta complicata refers to a clouding of the eye lens as a result of intraocular disease or eye surgery, caused by proliferation of lens epithelial cells on the posterior capsule. The most common are anterior uveitis without/with posterior synechia, myopia, and hereditary vitreoretinal diseases, such as retinopathy pigmentosa or untreated retinal detachment. The most common operations are filtrating glaucoma surgery or pars plana vitrectomy, which leads to clouding of the lens in 80–90% of patients in the first 2 years postoperatively.

Other causes of lens opacification occur after injuries (cat. traumatica). Opacities can occur due to blunt trauma (contusio bulbi) in the form of a contusion rosette (Fig. 4a) or opacities in the area of intraocular injuries due to fibrous metaplasia of the lens epithelium. In these cases, the epithelium can be stimulated to unspecific proliferation. These opacities may be partial or complete, and may be associated with swelling of the lens in various degrees with a dangerous increase in intraocular pressure. Occasionally, an intracapsular metallic foreign body can be observed

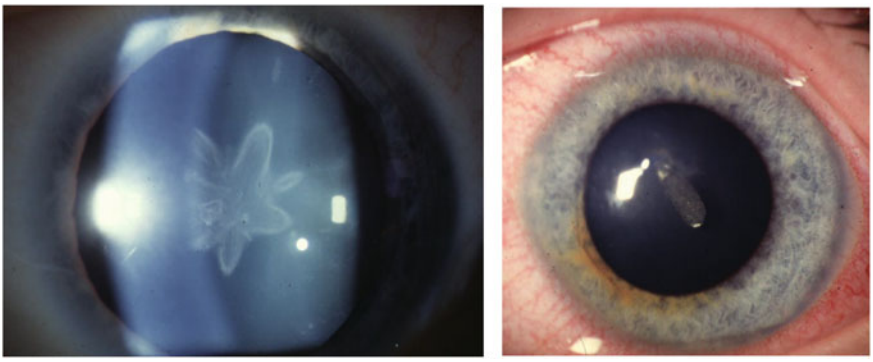

Fig. 4 Cat. traumatica with a contusion rosette (**a**) and intracapsular metallic foreign body (**b**)

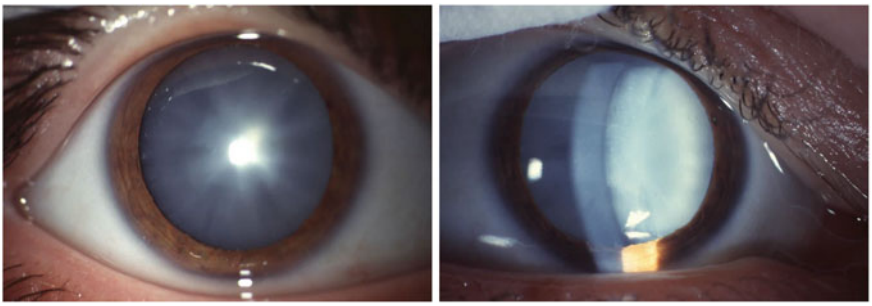

Fig. 5 Cat. electrica with complete opacity of the lens after a high-voltage accident while "surfing the suburban railway"

during slit-lamp examination (Fig. 4b). High-voltage accidents can also lead to a complete opacification of the lens (Fig. 5).

Other eye pathologies

Changes in the shape of the lens (coloboma, spherophakia, microphakia, lenticonus/lentiglobus) are usually due to embryonic malformations (see also Table 1). These can occur as an isolated anomaly or as a result of genetic defects (trisomy 13) or after intrauterine rubella infection in the first months of pregnancy.

Luxations of the lens

In the case of luxations or subluxations of the lens, a lens tremor (lentodonesis) and/or iris tremor (iridodonesis) is noticeable during slit-lamp examination with small eye movements. Typically, monocular double vision is caused by a displacement of the lens edge in the pupil. Causes of lens luxation include aplasias of the zonula fibres, which can be isolated or diffuse. Hereditary causes include Marfan syndrome (autosomal dominant) with typical segmental zonula defects inferiorly, leading to a superotemporal subluxation of the lens. Conversely, in homocystinuria (autosomal recessive/autosomal dominant) the progressive degeneration of the entire zonular apparatus due to cysteine inclusions leads to inferonasal luxation of the lens. Finally, in Weill-Marchesani syndrome (autosomal recessive, cystathionine synthetase decreased) we see a progressive degeneration and elongation of the zonular fibres with anterior and inferior dislocation, which is also associated with microspherophakia. Acquired causes include traumatic damage to the zonular fibres, and high myopia, usually due to diffuse zonular weakness. In the case of complete dislocation from its normal position, the lens may shift to the anterior chamber or drop into the vitreous.

Note

In disorders of the zonula fibers cataract surgery is significantly more difficult.

Pseudoexfoliation syndrome (PEX), is a generalised degenerative process of the extracellular matrix, and can also lead to instability of the suspension apparatus

through focal infiltration of the zonula fibres [1]. The presence of PEX at an older age can be easily detected at the slit lamp by the typical circular, whitish deposits of a fine fibrillar protein on the edge of the iris and anterior lens capsule.

Grading of Cataracts

A clinical assessment of lens opacity (grading) is important to document prior to surgery. This is usually carried out at the slit lamp but can be influenced by subjective bias from the examiner observations. In clinical trials, objective assessment methods are preferred to the examiner's own opinions to better classify the patient's symptoms of visual deterioration, glare and double vision.

Since cataracts cause opacity and increased light scattering resulting from disturbed fibre architecture and/or spatial arrangement of protein molecules in the fibre cells [1], a systematic assessment of lens opacity (lens opacity classification system, LOCS) was been proposed [4]. The LOCS-III system can be performed at the slit lamp or by using standardised photographs and has proven particularly useful for scientific comparisons [4, 5]. In this system, three forms of opacification are divided into 5–6 stages and subjectively assessed using a classification system. A distinction is made between opacification of the nucleus (nuclear), the cortex (cortical) and the posterior lens (subcapsular). In the case of nuclear opacities, the extent of the opacity (opalescence) and the colour intensity (colour) are also subdivided.

The further development of imaging techniques has also been useful in the assessment of lens opacity. Optical methods based on the Scheimpflug principle, aberrometry (double-pass technique) and optical coherence tomography (OCT) have all been applied to the problem of objective quantification of lens opacity.

Scheimpflug based imaging techniques use a blue-light emitting diode to display the anterior segments of the eye [6]. With the “Pentacam nucleus staging” (PNS) a cataract classification in steps from 0 (without opacity) to 5 (complete lens opacity) and a lens density from 0 to 100% have been described. A lens density greater than 11% and a PNS greater than 1 are regarded as limits for cataract classification.

Aberrometry involves projecting an infrared laser beam into the eye and recording the light scattering by a camera [7]. In this double-pass technique, an “ocular scatter index” (OSI) is determined, where a higher light scatter of >1 OSI indicates a cataract with a decrease in visual acuity.

Optical coherence tomography (OCT) imaging systems have been applied to examine lens density using both spectral-domain (SD) and swept-source (SS) techniques. Lens density was determined using the older SD-OCT but this has been superseded by automated lens densitometry using SS-OCT (IOL-Master 700, Zeiss) [8, 9]. With the “average lens densitometry” (ALD) a greyscale analysis of the image data points (pixels) can be performed. A higher intensity of the light reflections in the data points was associated with a higher cataract density. A value

of >74 units (pixel units) combined with a decrease in visual acuity showed a high sensitivity and specificity for cataract [9].

All methods were reproducible and reliable, but a good agreement with clinical cataract findings was not always seen. However, there was a clear improvement compared over the error-prone photo documentation. In general, the density measurement was more reliable, as the different colouring is not a good indicator of lens opacity, which has been clinically confirmed over time.

In summary, the clinical classification of lens opacity coupled with the typical symptoms is still the cornerstone for the preoperative examination of cataract. The standardised classification procedures (LOCS-III) or the newer imaging techniques can further add to the objectification of the cataract and can be useful in both clinical decision making and scientific comparisons. In the future, lens densitometry could be integrated into optical biometric devices for IOL calculation.

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