

Quick guide

Developmental prosopagnosia

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What is developmental prosopagnosia? Developmental prosopagnosia, also known as congenital prosopagnosia or ‘face blindness’, is a lifelong condition that affects individuals’ ability to recognise faces. Unlike cases of *acquired prosopagnosia*, where people encounter face recognition difficulties as an adult following a stroke or traffic accident, individuals with developmental prosopagnosia develop recognition problems in the absence of manifest brain injury. Individuals with developmental prosopagnosia experience difficulties recognising faces despite normal intelligence, memory, and typical low-level vision.

Which perceptual judgements are affected? Developmental prosopagnosics have difficulties recognising personally familiar faces. In severe cases, sufferers may find it hard to recognise partners and family members. However, deficits also extend to the perception of unfamiliar faces, for example, many developmental prosopagnosics have problems judging whether two unfamiliar faces presented sequentially are identical, particularly when external facial features (for example, ears, hairline) are occluded. As well as difficulties recognising and discriminating facial identities, some experience problems judging other facial attributes including expressions, gender, and attractiveness. In some cases, perceptual difficulties are limited to faces; other individuals, however, exhibit wider object recognition deficits that affect their ability to identify within-class exemplars (for example, identifying a particular bicycle within a set of bicycles).

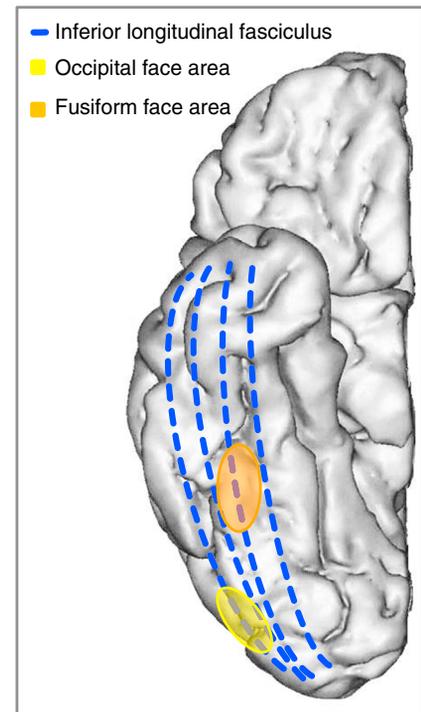
How do developmental prosopagnosics recognise others? Because of problems recognising people from their faces, individuals with developmental prosopagnosia learn to use alternative cues to recognise others.

For example, many sufferers recognise familiar others using characteristic facial features (such as an unusual nose), voices, hairstyles, clothing, and walking gait. Social situations where people wear similar clothes (such as school or work uniforms), or where voice cues are not available, frequently prove problematic for prosopagnosics. Similarly, familiar people are often misidentified when they change their hairstyle or wear hats.

How does developmental prosopagnosia affect the lives of sufferers? Developmental prosopagnosia can impact substantially on the lives of sufferers. At school, children with the condition can have problems recognising friends and teachers. As adults, some choose careers that do not require frequent face-to-face contact, and many avoid potentially challenging social situations. Problems following films and TV shows as a result of difficulties recognising characters in different scenes are also common. Often sufferers grow up blaming themselves, attributing their face recognition difficulties to attention deficits or poor memory. Some develop anxiety and depression as a result of the social difficulties they experience.

How prevalent is developmental prosopagnosia? Historically, cases of developmental prosopagnosia were thought to be extremely rare. However, considerable media interest has raised awareness in recent years and more sufferers have made themselves known to researchers. Current estimates suggest that as many as one in every 50 people might experience lifelong face recognition difficulties severe enough to interfere with their daily lives. If this estimate is accurate, the number of people affected might be approximately 1.5 million in a country the size of the United Kingdom.

How is developmental prosopagnosia identified? Identifying cases of developmental prosopagnosia is not straightforward. The condition is not listed in the Diagnostic and Statistical Manual of Mental Disorders (DSM-5) as a psychiatric disorder and no formal diagnostic criteria exist. Current approaches to diagnosis emphasise performance on objective, computer-based tests of face recognition ability,



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Figure 1. An illustration of the right hemisphere of the human brain viewed from below.

Reduced density and coherence of the inferior longitudinal fasciculus may impair information exchange between the occipital and fusiform face regions in developmental prosopagnosia.

including the Cambridge Face Memory Test (*participants are required to identify target faces presented alongside two distractor faces*) and the Cambridge Face Perception Test (*participants are required to re-order a sequence of faces according to their resemblance to a target face*). Leading research groups typically use several computer-based face recognition tasks and self-report instruments to develop a perceptual profile, and diagnose developmental prosopagnosia only where convergent evidence accumulates.

What is known about the origins of developmental prosopagnosia? The origin of the face recognition deficits seen in developmental prosopagnosia remains unclear. The condition often runs in families, suggestive of a genetic component. Cognitive accounts have argued that, relative to typically developing individuals, developmental prosopagnosics exhibit reduced holistic processing of

faces — whereby individual features are integrated into a coherent unified whole — compromising the accuracy and efficiency of their face recognition. Neuroimaging studies have revealed subtle neuroanatomical differences that accompany the condition. Several regions of the visual brain known to play an important role in face recognition, including the fusiform and occipital face areas, appear to be under-connected in developmental prosopagnosia, possibly impairing information exchange within this network. In particular, the integrity of the inferior longitudinal fasciculus, a white matter tract connecting the occipital and temporal lobes, is reduced in many developmental prosopagnosics (Figure 1).

Is developmental prosopagnosia related to autism? Developmental prosopagnosia may be more common in individuals with autism spectrum disorder than in the general population. Importantly, however, the two conditions are independent; many individuals develop autism spectrum disorder in the absence of face recognition difficulties, and many prosopagnosics exhibit no signs of autistic symptomology. Developmental prosopagnosia is an example of a ‘neurodevelopmental’ condition, similar to dyslexia, dyspraxia, dyscalculia, and autism spectrum disorder. Many neurodevelopmental disorders are known to co-occur. Genetic or environmental factors that cause an individual to develop a neurodevelopmental condition, such as developmental prosopagnosia, appear to increase their chances of developing others.

Where can I find out more?

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Quick guide
Bioluminescence

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What is bioluminescence?

Bioluminescence is the emission of light by an organism as a result of a biochemical reaction. In contrast to fluorescence and phosphorescence, bioluminescence reactions do not require the initial absorption of sunlight or other electromagnetic radiation by a molecule or pigment to emit light. Bioluminescent systems produce light through the oxygenation of a substrate, generically called luciferin (lat. *lucifer*, the light-bringer), and an enzyme, luciferase. Bioluminescent reactions vary greatly among organisms but can generally be described as a luciferase catalyzed production of an excited intermediate from oxygen and luciferin that emits light when returning to its ground state. Additionally, many bioluminescence systems involve cofactors such as FMN, ATP, additional enzymes and intermediate steps for the light production (Figure 1E). In some bioluminescence systems special types of luciferases, photoproteins, bind and stabilize the oxygenated luciferin and emit light only in the presence of cations, such as Mg²⁺ or Ca²⁺, which acts as a mechanism for the host to precisely control the timing of the light emission.

How does bioluminescent light differ among organisms? Light production in bioluminescence has a remarkable range of emission patterns such as continuous glow (Figure 1A,C), single flashes of light, e.g. in dinoflagellates, or repetitive pulse patterns that are often species-specific (Figure 1B). Bioluminescent light is emitted in wavelengths between 400 and 720 nm, from violet into the near-infrared. The majority of bioluminescent marine organisms emit blue light (410–550 nm), which correlates with the peak sensitivities of the opsins of many marine organisms. Interestingly, wavelengths of bioluminescent light seem to shift based on the habitat of the organism:

from violet and blue (420–500 nm) in the deep sea to blue-green (460–520 nm) in shallow waters to green-yellow (520–580 nm) on land, and its hue is often correlated with the optical characteristics of the environment (Figure 1D). The color of the bioluminescent light is dependent on multiple factors such as the luciferins and luciferases that are involved in the bioluminescent reaction or the conformation of the luciferase. In some bioluminescent systems fluorescent pigments, e.g. the green fluorescent protein (GFP), act as secondary emitters that affect the emitted color of the light. In addition to variations of the light-emitting molecules some organisms alter the original color of the bioluminescence through anatomical structures that act as biological filters and can refract or reflect the emitted light.

How is bioluminescence distributed among taxa? To date, bioluminescence has been reported in nearly 700 prokaryotic and eukaryotic genera. The majority of bioluminescent organisms inhabit marine environments including bacteria, dinoflagellates (the well-known ocean glow), molluscs, crustaceans, bony fish and sharks. In contrast to marine species, bioluminescence has not been confirmed in any fresh water organisms. On land, bioluminescence is less common and almost exclusively found in fungi and animals. Approximately 70 species of fungi in four lineages of the order Agaricales are bioluminescent. In animals, bioluminescence has been reported in two phyla, Nematoda and Arthropoda. Phylum Arthropoda includes one of the best-known groups of terrestrial bioluminescent organisms, the fireflies (order: Coleoptera).

How diverse are bioluminescence systems and what are their evolutionary origins? Bioluminescent systems are as diverse as their host organisms. Few bioluminescence systems are conserved among related taxa with the exception of fungi which are thought to share one conserved bioluminescence system. Generally, luciferins are more widely conserved than luciferases. For example, the

