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Congenital Horner Syndrome in art: the case of a historical Austro-Hungarian soldier

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Abstract. One of the most important characteristics, which determine facial appearance, is eye colour. Among the different conditions that causes an alteration in the normal iris pigmentation is congenital Horner Syndrome. Here we describe a case of right unilateral segmental heterochromia iridum in the historical portrait "Portrait of an unknown soldier" displayed at the Ferdinandeum Tyrolean State Museum (Innsbruck, Austria). Along with heterochromia, the sitter also displays right anisocoria, miosis, and mild ptosis of the upper and lower lids. This case highlights the interest of artists for modification of facial appearance in order to represent the sitter in the most possible realistic way.

Keywords: heterochromia iridum, anisocoria, miosis, ptosis, Renaissance, Austria.

Segmental heterochromia is rare in humans. The majority of cases of heterochromia iridum is benign and is not associated with underlying disorders. However, this condition needs monitoring, since it may be associated with different diseases, such as congenital Horner Syndrome, Parry-Romberg Syndrome, Waardenburg Syndrome and Fuchs heterochromic iridocyclitis (Rehaman, 2008).

Here we present a case of right unilateral segmental heterochromia iridum in the "Portrait of an unknown soldier" (Inventory number Gem 697) displayed at the Ferdinandeum Tyrolean State Museum (Innsbruck, Austria) (Figure 1). The painting has been prepared in the style of the school of Rubens. However technical investigations suggest that the portrait presents a

possibly 19th century repaint of an older canvas. Therefore, an exact dating of the painting is currently not possible. Along with heterochromia, the sitter also displays right anisocoria, miosis, and mild ptosis of the upper and lower lids. This triad of clinical features is consistent with congenital Horner syndrome. Facial anhidrosis, another feature which may be present in Horner Syndrome (HS), cannot be appreciated in a canvas.

First described in animals by Claude Bernard (1854), the syndrome was identified in humans in 1863 by Weir-Mitchell and colleagues. However, it was not until 1869 that the Swiss ophthalmologist Johann Friedrich Horner (1831-1886) attributed it to oculo-sympathetic paresis (Kanagalingam, 2015; Jeffery et al., 1998). The palsy manifests when lesions of one of the three neurons forming the oculosympathetic pathway (the central neuron, the preganglionic neuron, or postganglionic neuron) occur the pupil and the eyelids is interrupted (Kanagalingam & Miller, 2015; Jeffery et al. 1998).

Depending on the affected neuron, a first order HS, second order HS, or third order HS, is diagnosed (Kanagalingam & Miller, 2015; Jeffery et al., 1998). In general, the first and third order neurons are more often involved in adults whereas, in paediatric patients, the second order neuron is the most likely. The lighter coloured iris is always on the side of the HS since sympathetic stimulation is needed for the deposition of the pigment in the stroma of the iris (Pollard et al., 2010). Denervation impairs melanin pigmentation making the affected eye lighter (Ropper & Brown, 2005).

Heterochromia iridum typically is a feature of congenital HS, but it can also occur in lesions with onset before 2 years old (Jeffery et al., 1998; Pollard et al., 2010). The most frequent cause of paediatric acquired HS at the second order neuron is neuroblastoma of the paravertebral sympathetic chain (Pollard et al., 2010). Given the mature age of the sitter, the diagnosis of neuroblastoma can be excluded. Also Parry-Romberg Syndrome, Waardenburg Syndrome and Fuchs heterochromic iridocyciclitis (Rehaman, 2008) can be excluded, since their clinical features would have not been consistent with military life. Similarly, except from birth trauma or postviral infections, lesions due to other serious and life-threatening acquired conditions can be reasonably ruled out (Pollard et al., 2010).

All clinical features displayed in adult-onset acquired HS (i.e. Pancoast tumour, dissecting carotid aneurysm, middle cranial fossa neoplasm, brachial plexus trauma, brainstem stroke, carotid artery ischaemia and migraine) can be confidently excluded, since heterochromia iridum is not manifested in these conditions (Kanagalingam & Miller, 2015; Jeffery et al., 1998; Pollard et al., 2010). Furthermore, in adults with HS, acquired heterochromia is extremely rare. So far, the disorder has never been reported in patients with an acute or recently acquired HS (Diesenhouse et al., 1992).and only a few cases of long-standing, acquired Horner Syndrome with heterochromia iridis are mentioned in biomedical literature (Diesenhouse et al., 1992).

Figure 1. Portrait of an unknown soldier, oil on oak wood, 72,5 x 55,4 cm. On the rear side an Anvers burn mark. Inventory number Gem 697. Legat [bequest] Josef Tschager 1856 (provenience: Collection Ritter von Prohaska, auction Mollo (Vienna) 12. March 1820). While the profession of the sitter was inferred from the military uniform, no further personal information is available. The canvas is attributed to the school of Rubens and has been tentatively dated between 1577-1640. The canvas is currently held by the Ferdinandeum, the third oldest national Museum of the Austro-Hungarian Empire, which was founded in 1823 and named after Archduke Ferdinand II.

Taking in account that the unknown sitter was a soldier, the presence of an artificial right eye should be considered. However, although the use of eye prosthetics dates back to Antiquity (up to 4800 years ago) (Pine et al., 2015; Moghadasi, 2014); the ability in manufacturing eye prosthetics reached high technological levels in the late 16th century with Venetian glass blowers, along with lens makers, able to produce realistic but fragile glass eyes (Pine et al., 2015). High quality glass eyes were also

produced in Augsburg where a manufacturing centre was active until the Thirty Years War (1618-1648) (Pine et al., 2015). However, based on the pictorial representation, eye trauma is very unlikely, since no scarring in the eyelids can be appreciated. This absence of evidence leads us to exclude the presence of a glass eye.

This pictorial representation of congenital HS adds to another we previously identified in a portrait displayed in Vienna's *Kunsthistorisches Museum* (Austria) (Bianucci et al., 2020). In that canvas, which originally belonged to the pictorial collection of Archduke Ferdinand II of Tyrol (1529–1595), a sitter, another unknown adult nobleman, appears to be affected by paediatric HS in the right eye. These cases highlight the peculiar interest of both the artists and their patrons to depict physical modification from the normally perceived anatomy of the eyes and ocular eyelids. In addition, these examples strongly indicate that (at that time) individual traits, including pathological changes, were documented accurately raising the value of the paintings as realistic depictions.

The main limitation that needs to be emphasised is the lack of contemporaneous documentary sources and remains of this individual to triangulate the findings. However, the portrait nonetheless offers a visual image that gives insight into the past presentation of a relevant medical condition.

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