Diplophthalmia versus cyclopia and synophthalmia
Mechanisms of doubling of the eye

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Abstract. In historical and many scientific papers of the 20th century double formation of eyes has been described as cyclopia, which develops as a variably marked fusion of both eye anlagen into a single eye in median position. In contrast with mythology, no cases were viable, as cyclopia is always accompanied by other severe deformities, such as cerebral defects and proboscis. For the first time a double eye on one side - a unilateral diplophthalmia - in a viable child born in Austria is presented after a reviewal of fundamentals of general and experimental embryology, cyclopia and unilateral synophthalmia. Unlike cases of cyclopia with two fused eyes from two anlagen in one single orbit, we found a total of three eyes from two anlagen in two orbits; a normally developed eye in the right orbit and a double eye in the left with no proboscis. In addition porencephaly, a double canine tooth, and a lateral branchial cyst were also present on the left side. An examination of this unique case partially corroborated the findings of experimental embryology as to the normal formation of the lens and brought to light new findings concerning the formation of the vitreous body and the retina, because in the 'extra' left eye a hyaloid artery did not develop. There was only one optic nerve with a normal optic disc as a guiding structure for this artery in the 'normal' left eye.

1. History and general background

Cyclopia and associated malformations are the most grotesque developmental abnormalities in ophthalmology. Malformed human monsters with a single median eye have been reported for several thousand years. Cuneiform characters on Babylonian clay tablets refer to such a monster bringing calamity to the country. These tablets were found near the Tigris River and may be almost 4000 years old [1]. Other mythological examples are found as Polyphem in Homer's *Odyssey*, 800 BC, as three cyclopic storm gods in Hesiod's *Theogony*, 700 BC and in Euripides's *Cyclops*, 410 BC. Then literature was rich in allusions to that picturesque, though imaginary race. Descriptions of cyclopean monsters are not uncommon in papers of the natural philosophers of the 16th and 17th centuries. They are, however, typically inaccurate and often fanciful, e.g., the cyclopean woman from Reisch's *Margarita Philosophica* of 1508 [2] (Fig. 1) and the cyclopean infant
Fig. 1. The cyclopean woman (from Reisch's *Margarita Philosophica*, 1508).

Fig. 2. The cyclopean pig, painted by Carus 1842 [4]. Note the still fanciful presentation, but the unconscious presentation of the later ‘twins theory’.