

*Citation:*

Winkler, C., On Cyclopa with conservation of the Rhinencephalon, in:  
KNAW, Proceedings, 22 I, 1919-1920, Amsterdam, 1919, pp. 283-290

**Physiology.** — “*On Cyclopia with conservation of the Rhinencephalon*”. By Prof. C. WINKLER.

(Communicated in the meeting of May 31, 1919).

In These Proceedings of February 1916 I reported the results found after examination of the brains of three cycloopian monstra.

I then pointed out, that the characteristic abnormality in all these brains, was the presence of a sack with a thin wall, formed by the roof of the third ventricle, largely extended by fluid.

I was forced to contradict a sentence, found in SCHWALBE's “*Die Morphologie der Missbildungen*” where he says: “*das Vorderhirn ist bei den Cyclopien nie in Hemisphären geteilt*”. I must assert, that in all cases of cycloopian brain the hemispheres are well differentiated at the occipital end.

At the frontal pole however they are often united, although in one of the cases then described, there was also a sagittal fissure at the frontal pole of the hemisphere.

I therefore deny that the examination of cycloopian brains should give any support to the supposition that the terminating-time of the cyclopia must be placed in a period, preceding that in which the sagittal fissure of the telencephalon appears. Also the view that cyclopia is *inevitably* accompanied by arhinencephalia, I could only accept under certain reserve.

In fact, in all the cases which I examined, the bulbi olfactorii and the lobi olfactorii anteriores were missing. But the lobi olfactorii posteriores were present. They are found along the mesial line of the brain-base. There they were placed next to each other, because the brain-base between them was missing.

Ordinarily cycloopian brains are partially arhinencephalic, missing only the frontal part of the rhinencephalon.

Since then I have prepared the brains of several cases of cyclopia, largely differing between each other.

Now I believe that there is another monstrosity, the so-called synotia, nearly related to the cycloopian one. The former is often combined with the latter.

Through the kind cooperation of Professor KOUWER and Dr. VERMEULEN I obtained possession of such-like monstrosities.

1. a human fetus with synotia (nose, eyes and maxilla superior are existing). There is an indication of a mouth. No inferior maxilla is found. The ears are united in the mid-line in a single meatus acusticus externus (In this being the brains were unfortunately thrown away).

2. a fetus of a pig with cebocephalia (Proboscis is found above two totally separated eyes in one orbit).

3. a fetus of a lamb with incomplete cyclopia (one oblong eye with a long pupil, two optic nerves; no proboscis is visible by the naked eye, although the X-ray photo shows a little nasal bone at the os frontale).

4. a fetus of a calf with almost complete cyclopia (one oblong eye with hour-glass like pupil, two optic nerves closely situated, no proboscis.)

5. a fetus of a calf with incomplete cyclopia and synotia (two united eyes in the circle of the four eyelids, no proboscis. A mouth-opening, bordered at the upper part by a strong upper-jaw (X-ray photo). No lower jaw (X-ray photo), no tongue. The os hyoideum is completely developed (X-ray photo). The ears are united in the mid-line).

6. a fetus of a lamb with synotia. (A small proboscis, no eyes, no jaws, no mouth or tongue. There is an os hyoideum. (X-ray photo.) The animal therefore is anophthalmic, agnathic, aprosopic. The ears are united in the mid-line. Larynx and pharynx end towards the top in a blind sack. No thyroid gland.)

7. a fetus of a lamb with cyclopia and synotia, dealt with in this communication.

In all the brains of those monstra which were examined, there was found a membranous sack, which shows itself to be the roof of the third ventricle, much extended by fluid. Now and then however peculiarities were seen in this sack at its frontal or its occipital end. In No. 5 and No. 6 e.g. the sack was continued in the like-wise thin wall of the telencephalon and in N°. 7 at its distal end the mesencephalon and the cerebellum were not developed, forming a part of the thin wall of the sack covering also the IV ventricle.

The sack is not only existing in the brains of cyclopiian monstra, but also in those of the synotic type.

In another paper I will describe more in details the differences of the brains in those monstra.

Here I intend to demonstrate the monstrosity, mentioned sub N°. 7. It may be considered as a sample of a cyclopiian malformation, with

synotia possessing a complete rhinencephalon and therefore it already warns us that we should not rashly assert that every cyclopia must be arhinencephalic.

Regarding the drawing of this fetus, seen in the face, we notice

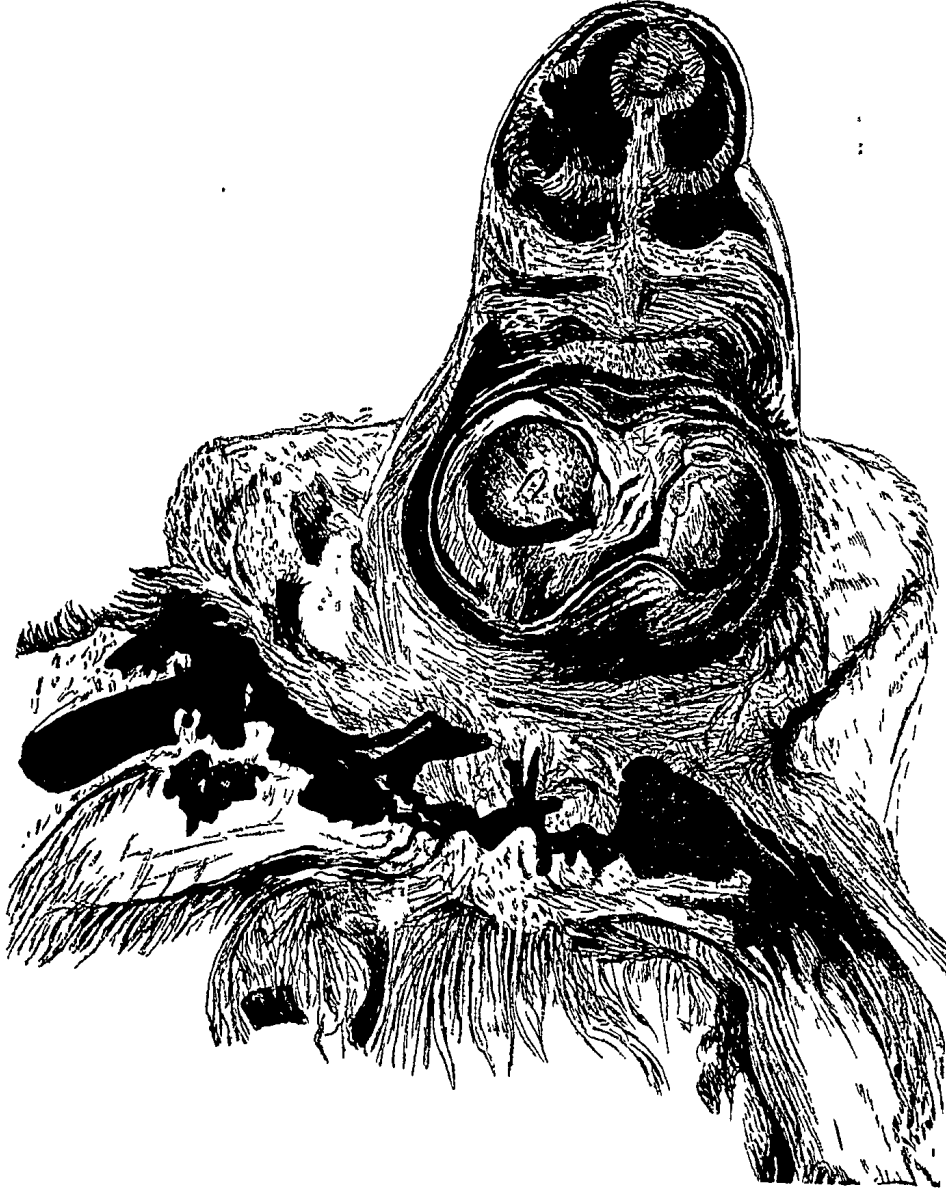


Fig. 1.

Drawing after a photograph of a lamb fetus with cebo-cephalia and synotia. The monstrosity possesses a well-shaped and completely developed nose above the two eyes, surrounded by four eye-lids. One can discover the two medial canthi. The ears are united together. No mouth.

that the animal possesses a complete nose with nostrils. Underneath it is found, that what appears to be an orbit, surrounded by four

eyelids and in which the two united eyes are placed with a smaller left and a larger right eye. Jaws, mouth and tongue are missing. Directly underneath the eyes the two ears lie by the side of each other in the mid-line and the two external auditory canals possess a combined opening.

Pharynx and larynx are well developed and continue in a hollow, ending upwards blind.

The X-ray photo confirms that the jaws are totally missing, but it also shows that the os hyoideum is well developed. Moreover the X-ray photo teaches that the ethmoidal bone is fully present. Crista

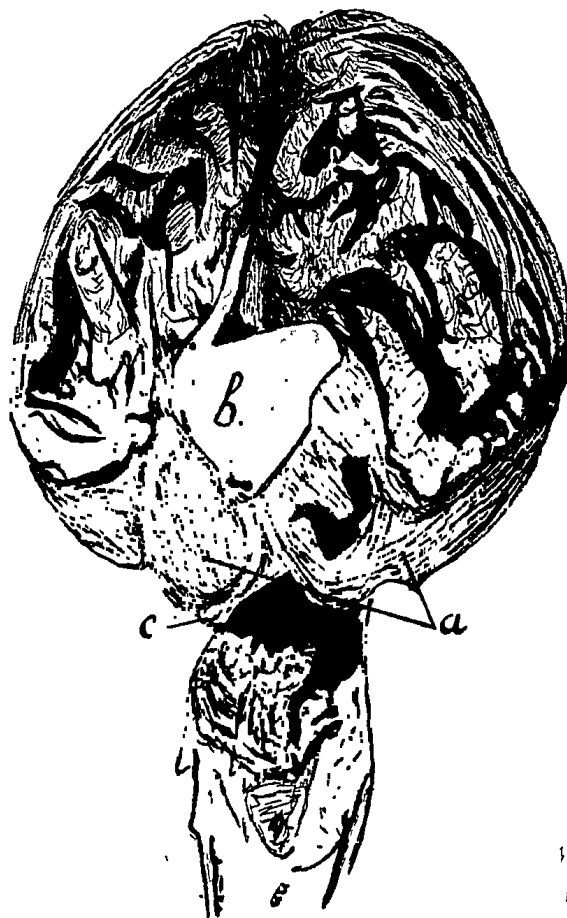


Fig. 2.

Drawing of the upper surface of the brains of the monstrosity reproduced in fig. 1. The sack (*a*) is visible. It adheres to the dura mater (*b*). At the distal end (*c*) it continues in the mesencephalon and in the cerebellum, represented by a thin membrane. Through this membrane shines the tela chorioidea of the fourth ventricle.

Galli, lamina cribrosa, lamina papyracea and the labyrinth of the ethmoidal bone can be distinguished. Already before removing the brains I therefore knew, that this cyclopic being could not have

been formed through the missing of the ethmoidal bone and of the mesial wall of the orbit.

On removal of the brains one is directly struck by the presence of the sack (fig. 2*b*) and the epiphysis lying free, seeming to be its point of origin. Behind it one does not find anything of a mesencephalon or of a cerebellum. The sack continues in occipital direction and the tela of the 4<sup>th</sup> ventricle shines faintly through it (fig. 2*c*). On the other hand both the hemispheres are well developed. There is a deep sagittal fissure, in which the dural septum with its sinus are found, and which has to be cut away from the crista galli in order to make its removal possible.

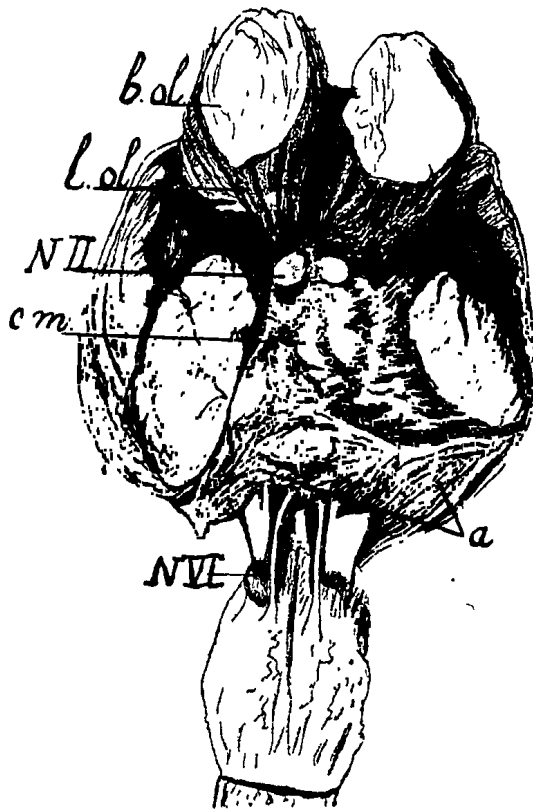


Fig. 3.

Drawing of the basal surface of the monstrosity reproduced in fig. 1. *a* = the thin membrane covering the defect in the pes pedunculi cerebri. *b. ol.* = bulbus olfactorius. *l. ol.* = lobus olfactorius. *N. II* = nervi optici *c. m.* = corpus mammillare, shining through the membrane. *N. VI* = nervus abducens.

If we examine the basal surface of the brain (fig. 3), then we are struck by the presence of two well-developed olfactory bulbs with their tract, their lobus olfactorius anterior, posterior and cornu Ammonis, in short of a completely developed rhinencephalon.

There are two *N. optici* (fig. 3, *N. II*). The corpus mammillare

shines through the pia mater. One can surely demonstrate the N. abducens of the cranial nerves. With the nerve lying proximally from this, it is not so. Only after a microscopic examination, it is certain that it is the N. trigeminus. Between these two thick nerves the base of the brain is formed by a thin membrane, which when the brain was removed could only be spared with the utmost care.

If the sack is opened at its dorsal part and is folded backward, there appears a local defect in the brain-base, more than 1 cm. wide. There the base of the brain is formed by a membrane, 1 mm. thick, at the utmost, in which some white nerve strings diverge from the mid-line towards a proximally placed mass of nervous tissue. This nervous mass, striatum and thalamencephalon are shining through the pia mater at the base in fig. 3. There is an interrupted continuation in the brain-base at the level of the pes pedunculi.

The bony base of the brain is very remarkable. The crista galli protrudes. On both its sides the lamina cribrosa carries the bulbi olfactorii, which send their fila olfactoria through it. Moreover the optic foramina are normally formed, together with the frontal part of the os sphenoidale. Then however the sella turcica is found missing, also the hypophysis. The base of the crane is not massive, but movable, as there is a great loss of bone distally from the sella turcica.

An X-ray photo taken from the upper side makes this obvious. Here a large defect in the bony base of the skull appears. The caudal part of the os sphenoidale is missing in the mid-line and the frontal part of the clivus has fallen out as far as the arch of the Atlas. The os petrosum is intact on both sides. There are no jaws. Through the loss of the facies orbitalis of the upper jaw the eyes have sunk downwards. They are no longer lying in a bony orbit. The lamina papyracea of the ethmoidal bone is placed proximally from the double eye and therefore easily recognisable in the X-ray photo. In this case, it is not because the mesial wall of the orbit has been destroyed that the eyes have met one another in the mid-line, but because the lower wall of the orbit is absent.

This cyclopic monstrosity possesses a complete rhinencephalon, but at the same time it becomes synotic through the loss of a lower jaw.

As to the question, what may be the cause of such a monstrosity one must acknowledge that immense difficulties arise in defending that there was an insufficiency of germ material, as far as the brain as well as the bony parts of the skull. is concerned. Simpler is the view in taking the sack as point of origin.

A pathological process which calls forth the sack (and the latter is found in all the cases of this sort) is able to destroy at the dorsal brainside the mesencephalon and the cerebellum when they hinder its development at the distal end. But at the ventral side the pathological process destroys the pedunculi cerebri and the tissue on which they lie. This is the germ material out of which jaws, mouth and tongue are going to develop.

Distally bordered by the second branchial arch (the os hyoideum was intact in all the three cases of synotia) all is destroyed that is going to develop out of the first branchial arch (except occasionally of the ossicula acustica) and out of the tissue, which lies proximally from it. In this way the local defect of the pedunculi cerebri and of the tissue forming the middle part of the skull, is easily understood.

According to the view which I explained in my previous report, I think the cause of the sack to be a local process of inflammation, which by means of a mechanical influence produces a defect at the base of the brain and of the skull.

On the other hand I acknowledge the possibility that the sack and the basal defect, together may be co-effects of another more complicated pathological cause.

In debates upon this subject, held at Leiden Dr. MURK JANSSEN defended the thesis that the narrowness of the amnion may perhaps produce the sack as well as the defect, by compressing the head of the embryo in a strictly defined plane. The result may be that all the germ material, which is found in this plane, may die. Should such a hypothesis be confirmed, there will arise different possibilities in the formation of these monstra, but I will not yet enter upon these.

I only wish to lay stress on the following views.

Destroying of tissue at the proximal end of the skull, so that the os ethmoidale disappears and the dorsally placed sack, the roof of the distended III<sup>rd</sup> ventricle is formed, gives rise to cyclopidian monstra. But they are not the only defect-formations which are found.

There is yet another place of predilection, where the tissue that will form parts of the skull, may be destroyed. In such cases the first branchial arch may be destroyed by pathological processes. These lead to synotia. Now and then it occurs isolated. Then we see uncomplicated synotia. Nose, eyes and upper jaw are well developed, as found in the fetus, mentioned sub I.

But also the two local destroying processes appear, independent of each other, next to one another. Then cyclopidian and synotian deformities are found together. And there may remain between



them an intact upper jaw as was the case with the monstrum, mentioned under 5.

Now and then the two local destroying processes are united. Then comes a massive fissure and the monstrosities appear, as described under 4 i.e. synotia with anophthalmia, aprosopia and agnatia.

The series of the different cyclopic deformities is joined to that of the synotian deformities. In this series one case is most remarkable, i.e. if the local destroying process, commencing at the first branchial arch spreads so far proximalwards that the upper jaw totally falls out.

Then the eyes are going to meet each other in the mid-line, while the maxillar part of the orbit is lost. Then also two eyes are found in a four eye-lid ring, but they are no longer placed in an orbit. In that case there is found a cebocephalic form of cyclopia, with a complete nose above the eyes, and a complete rhinencephalon.

To me it seems even possible that the local destroying process may spare the first branchial arch, only destroying the upper jaw. In such a case there results a cyclopia, perhaps always the cebocephalic form, but without loss of the ethmoid bone, with a complete rhinencephalon and without synotia.

---