

Holoprosencephaly (HPE) is the failure of the prosencephalon, or forebrain, to develop normally. The forebrain is a region of the brain in the fetus that develops into parts of the adult brain, including the cerebral cortex. Instead of the normal complete separation of the left and right halves of the forebrain, there is an abnormal continuity between the two sides. There are several different types of holoprosencephaly. In the alobar form, there is no separation between the right and left halves at all. In semilobar HPE, at least some separation of the two halves is present. In the lobar form, most of the brain has separated into right and left sides, though there is incomplete division into the two halves. Holoprosencephaly affects males and females in equal numbers before birth and has been reported in many ethnic groups. The incidence of holoprosencephaly has been estimated at 1 in 250 during early embryonic development, and approximately 1 in 16,000 live births.