Central Nervous System

Anencephaly
(Core Condition)

Description	Partial or complete absence of the brain and skull.



Inclusions	Acrania – Absence of skull bones with some brain tissue present. Absent brain, with or without skull bones present. Anencephaly Anencephaly Craniorachischisis – Anencephaly continuous with an open posterior spinal defect with no meninges covering the neural tissue. Exencephaly
Exclusions	Encephalocele Iniencephaly Rachischisis – When used alone, this term refers only to the spinal defect and should be coded as spina bifida without anencephaly.
ICD-9-CM Codes	740.0 – 740.1
ICD-10-CM Codes	Q00.0 - Q00.1
CDC/BPA Codes	740.00 – 740.10
Diagnostic Methods	Anencephaly is easily recognized on physical examination at delivery.
Prenatal Diagnoses Not Confirmed Postnatally	Anencephaly may be included when only diagnosed prenatally. However, if it is possible to ascertain the degree of certainty of the prenatal diagnosis, this should factor into the decision as to whether or not to include an individual case in the surveillance data.

Additional Information:
Anencephaly is one of a group of defects that result from failure of the neural tube to close.

Maternal serum alphafetoprotein (MSAFP) and/or amniotic fluid alphafetoprotein (AFAFP) and amniotic fluid acetylcholinesterase (ACHE) may be elevated with anencephaly. However, these screening tests alone are not sufficient to diagnose the condition.

In cases where both anencephaly and spina bifida are present but are not continuous (i.e., not craniorachischisis), both anencephaly and spina bifida should be coded.