Total Anomalous Pulmonary Venous Connection (TAPVC)

(Core Condition)

Description A condition in which all 4 pulmonary veins

connect anomalously into the systemic venous circulation to the right atrium or the body (systemic veins) instead of the left atrium; often occurs with other cardiac defects.



Inclusions TAPVC (total anomalous pulmonary venous connection)

TAPVR (total anomalous pulmonary venous return)
TAPVD (total anomalous pulmonary venous drainage)

Exclusions If not all 4 veins are visibly connecting/draining anomalously (e.g. Partial

Anomalous Venous Return, ICD-9-CM code 747.42 or CDC/BPA code

747.41 or Q26.3)

ICD-9-CM Codes 747.41

ICD-10-CM Codes Q26.2

CDC/BPA Codes 747.42

Diagnostic Methods While TAPVR may be suspected by clinical presentation, it may be

conclusively diagnosed only through direct visualization of the heart by cardiac echo (echocardiography), catheterization, surgery, or autopsy. The difficulty in viewing all 4 veins may mean that several echocardiograms may

be needed to confirm the diagnosis.

Prenatal Diagnoses Not Confirmed Postnatally TAPVR is difficult to identify prenatally. If identified by prenatal ultrasound, it should not be included in surveillance data without postnatal confirmation.

In addition, the absence of TAPVR on prenatal ultrasound does not

necessarily mean that it will not be diagnosed after delivery.

Additional Information:

Total anomalous pulmonary venous return and partial anomalous pulmonary venous return have not been shown to be developmentally related, although they share a similar description. Also, there are subtle differences in the meaning of anomalous venous connection, return, and drainage, but the terms are often used interchangeably.