



Clinical Practice Guideline for Care Around Stillbirth and Neonatal Death

Section 7 The PSANZ Classification System for Stillbirths and Neonatal Deaths

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Endorsed by













SECTION 7

THE PSANZ CLASSIFICATION SYSTEM FOR STILLBIRTHS AND NEONATAL DEATHS

7.1 Introduction

This section of the Perinatal Society of Australia and New Zealand (PSANZ) Clinical Practice Guideline for Care Around Stillbirth and Neonatal Death presents the third revision of the PSANZ Classification System for causes of stillbirths and neonatal deaths. The system was first released in May 2003.

Accurate classification of the causes of the stillbirths and neonatal deaths is the cornerstone of prevention and the ability to compare causes of death across and within countries is key to this effort. The main driver for the PSANZ system is the limitations of the International Classification of Disease (ICD) for classifying perinatal deaths¹⁻³. This has also been the case globally, with over 80 disparate systems recently identified, none of which appear optimal³. The new adaptation of ICD 10 by WHO – ICD for perinatal mortality (ICD-PM)⁴, which is recommended for use as part of perinatal mortality audit, holds promise for consistent global reporting of causes of perinatal deaths. Initial piloting of ICD-PM has highlighted important areas for improvement when ICD is next updated ⁵. The ICD-PM uses ICD rules based only on death certificate data⁴ and classifies the underlying cause of perinatal death. In Australia, death certificate reporting of causes of perinatal deaths (particularly stillbirths) is often inaccurate and overestimates the proportion of unexplained up to 50%⁶. This inaccuracy is partly a systems issue in that the mandated timing of completion of the death certificate precedes results of investigations, including autopsy, becoming available and also the lack of supervision and training in completion.

Until further enhancements are made to the ICD system, the PSANZ Classification System for Stillbirths and Neonatal deaths is recommended as the primary system for causes of perinatal deaths in Australian and New Zealand (ANZ). However, in order to facilitate global comparisons and to inform future improvements to ICD-PM, simultaneous application of ICD-PM and the PSANZ system or later mapping from PSANZ to ICD-PM is optimal. The ICD-PM (as for the PSANZ system) should be based on the findings of adequate investigation into the causes of death including committee review of the death (see Section 2 of the PSANZ Clinical Practice Guideline for Care Around Stillbirth and Neonatal Death).

The PSANZ system has been shown to perform well against other systems^{3,7}. In a recent evaluation of global classification systems for perinatal deaths against expert consensus of a quality system, some limitations were identified including the need for better definitions and rules. In this update, an attempt has been made to address these limitations. Accurate classification of stillbirths and neonatal deaths is only possible following adequate investigation and review of the clinical circumstances of the death and is an integral part of high quality perinatal mortality audit.

The standardised perinatal mortality data collection forms for New Zealand (see Appendix F – New Zealand rapid reporting form for a perinatal death – baby and Appendix G – New Zealand rapid reporting form for a perinatal death – mother) and for Australia (The Australian Perinatal Mortality Audit Tool (see Appendix E – Australian perinatal mortality audit tool) should be completed by the hospital perinatal committees to enhance the quality of classification of death.





For a history on the development of the PSANZ PDC and a summary of the changes made to the system since its inception please refer to *Appendix V – Development of PSANZ Perinatal Death Classification and PSANZ Neonatal Death Classification*.

This update of the guideline has been undertaken through a partnership between the PSANZ Stillbirth and Neonatal Death Alliance and the NHMRC Stillbirth Centre of Research Excellence in Stillbirth.

7.2 Principles, structure and performance indicators

Principles

The key principles of the PSANZ system are:

- To identify an underlying cause of death for stillbirths and neonatal deaths
- To identify up to two associated conditions for stillbirths and neonatal deaths
- To enable reporting by ICD-PM through identifying timing of death and mapping of categories to ICD-PM.

Including the assigned PSANZ system category codes as part of routinely collected individual birth record data across ANZ jurisdictions will enable reporting by timing of death (antepartum, intrapartum, early and late neonatal deaths or timing of death unknown) and also allows more detailed analyses by demographic and clinical factors to aid identify where attention is most needed.

Structure

The PSANZ System for stillbirths and neonatal deaths consists of two main sets of conditions (categories) and one set of associated conditions (contributory).

The two main category grouping are: 1) The Perinatal Death Classification (PDC) which includes maternal/fetal causes of stillbirths and neonatal deaths; and 2) The Neonatal Death Classification (NDC) including neonatal causes of death.

The PSANZ Associated Conditions list includes both the PDC and NDC categories plus other associated factors e.g. maternal risk factors and placental pathologies.

Performance indicators

Evaluation of the PSANZ system is being planned including the following measures of success:

- 10% or less deaths classified as Other unspecified
- 20% or less stillbirths classified as *Unexplained* with full investigation
- Ease of use by clinicians
- Valued by end-users
- Good to excellent agreement between classifiers assigned within the major categories
- Good alignment with ICD-PM

7.3 Objective of this section

The main objective of section 7 of the PSANZ Clinical Practice Guideline for Care Around Stillbirth and Neonatal Death is to present the PSANZ system for stillbirths and neonatal deaths and to provide detailed instructions for use to ensure consistent and comprehensive data on the underlying causes and associated conditions of stillbirths and neonatal deaths across Australia and New Zealand.





7.4 What has changed in this update?

In this update, changes have been made to categories based on new knowledge around the causes of perinatal death and to address deficiencies identified in a recent evaluation⁸.

The PSANZ system no longer uses a hierarchical approach between categories but rather employs rules around common scenarios when multiple factors are involved. This is to ensure consistency with the ICD principles of identifying an underlying cause of death.

Improvements in classification of placental pathology have been made although limitations remain and further research and consistency with reporting are needed to better define placental categories⁹. The 'Unexplained Antepartum Stillbirth' category now excludes stillbirths as a result of placental insufficiency and identifies those which were inadequately investigated. Placental pathology is now identified in more detail as Category 9 Placental dysfunction, replacing the previous category of 'Fetal Growth Restriction' (FGR). If present, FGR is now classified as an associated condition. The list of associated conditions has been expanded to include placental pathology thought to be contributory but not causal. Congenital anomalies have been updated to include more detail on chromosome and genetic conditions in line with advances in prenatal testing, and to align categories with *ICD-10 version 2016 Chapter XVII Congenital malformations, deformations and chromosomal abnormalities* (http://apps.who.int/classifications/icd10/browse/2016/en#/XVII).

The subcategories on the duration of membrane rupture (MR) in Category 10 Spontaneous preterm have been removed and a subcategory to identify cervical shortening preceding MR has been added. The Congenital anomaly category (Category 1) has been revised to acknowledge developments in genetic testing. Lastly, a new category (Category 6) to capture complications of multiple pregnancy.

The PSANZ Associated Conditions list includes both the PDC and NDC categories plus other associated factors e.g. maternal risk factors and placental pathologies.

As with other sections of this guideline, the formatting has been revised to enhance readability and reduce duplication across the different sections of the guideline. Please refer to Appendix U – Changes on this version of the classifications system to see a full list of changes in this revision.



Section 7 Recommendations for all perinatal deaths

- All stillbirths and neonatal deaths should be classified according to the PSANZ classification system to identify a single underlying cause of death for both stillbirths and neonatal deaths and up to two associated factors which contributed to the death.
- The classification of stillbirths and neonatal deaths should be based on the best available information from a comprehensive history and appropriate investigation (as recommended in Sections 4 and 5 of this guideline) and should form part of a formal institutional clinical audit process as outlined in Section 2 of this guideline.
- The classification should be included in the routine perinatal data collections across jurisdictions for every perinatal death to enable comprehensive reporting regionally and nationally including disaggregation and identification of timing of the death (i.e. antepartum, intrapartum, early and late neonatal deaths)
- 4 Following application of the PSANZ system, mapping to ICD-PM categories should be undertaken to enable high quality global reporting. This will often require alteration to the cause of death on the perinatal death certificate.

7.5 Purpose of the PSANZ System

The purpose of the PSANZ Perinatal Death Classification System is to ensure comprehensive and consistent data on causes and associated conditions for stillbirths and neonatal deaths across Australia and New Zealand to enable benchmarking and monitoring of causes of death to inform policy, practice and research, to help parents understand why the death occurred and to assist in future pregnancy planning.

7.6 General rules for applying the PSANZ PDC System

Classification of underlying cause and associated conditions

In accordance with ICD-PM, the PSANZ Classification System identifies a single underlying cause of death for stillbirths and neonatal deaths as well as the presence of associated conditions. For all stillbirths and neonatal deaths a maternal/fetal condition according to the PDC is assigned and, in addition for neonatal deaths, the underlying neonatal condition which caused the death is assigned according to the NDC. Therefore, for neonatal deaths the PSANZ system at least two conditions are assigned; the neonatal condition which resulted in the death and a maternal/fetal condition (according to the PDC). If no maternal/fetal condition is identified the classification category of "no obstetric antecedent" is applied.

Definitions

Underlying cause of death: According to ICD

"the disease or injury which initiated the train of morbid events leading directly to a person's death or the circumstances of the accident or violence which produced the fatal injury, as represented by a code¹⁰"





Associated conditions are defined as conditions which were considered to have contributed to the death but are not considered to be the main underlying cause. Conditions which were present but not considered to be contributory are not classified as associated conditions.

Please refer to the PSANZ Associated Conditions list (see page 35).

Classification of terminations of pregnancy

All terminations of pregnancy are identified by the inclusion of an "009" for two-digit codes and a "09" for the three digit codes and "9" for four digit codes i.e. 1.1 becomes 1.1009; 1.83 becomes 1.8309; 6.111 becomes 6.1119. This includes induction of labour without expectation of fetal survival e.g. in the case of severe pre-eclampsia at pre-viable gestations, or prolonged premature rupture of membranes with severe infection.

Classification numbering approach

As far as possible, the subcategory .8 has been used for 'Other conditions' and .9 for 'Unspecified conditions' within its category, as has been the case in the ICD classification.

If data are entered with a decimal point, a subcategory such as 'Structural anomaly' (Category 1, *Congenital Anomaly*) would be 1.1, but as a 4 digit numeric would be 0110. Similarly subcategory 'Group B Streptococcus' (Category 2, *Perinatal Infection*) would be 2.11 or 0211.

Inclusion in routine perinatal data collections at the individual case record level

It is recommended that PSANZ classification codes are included within routine perinatal data collections in each region for every perinatal death to enable disaggregation to better identify areas to focus prevention e.g. by Indigenous and socioeconomic status and other risk factors, and timing of death. The ability to analyse causes of perinatal deaths by timing of death (i.e. antepartum, intrapartum, neonatal, or unknown) is consistent with ICD-PM rules.

Reporting according to ICD-PM

Reporting by ICD-PM system enables international comparisons and should be based on the causes of perinatal deaths following thorough investigation and perinatal mortality committee review. Following application of the PSANZ classification system to stillbirths and neonatal deaths, mapping of the categories to ICD-PM should be undertaken for global reporting requirements. Jurisdictions may wish to classify according to ICD-PM simultaneously with the PSANZ system to assist in global reporting and to inform future improvements in classification.





7.7 PSANZ-PDC Classification including rules and definitions

1 Congenital anomaly

- 1.1 Structural anomaly
 - 1.11 Nervous system
 - 1.12 Cardiovascular system
 - 1.13 Genitourinary system
 - 1.14 Gastrointestinal system
 - 1.15 Musculoskeletal
 - 1.151 Congenital diaphragmatic hernia
 - 1.152 Gastroschisis/omphalocele
 - 1.16 Respiratory system (include congenital pulmonary airway malformation (CPAM))
 - 1.17 Haematological
 - 1.18 Multiple Congenital anomaly (no chromosomal/genetic cause or not tested)
 - 1.19 Other congenital abnormality
 - 1.192 Idiopathic hydrops fetalis
 - 1.193 Fetal tumour (include sacro-coccygeal teratoma)
 - 1.198 Other specified
 - 1.199 Congenital anomaly, unspecified
- 1.2 Chromosomal anomaly
 - 1.21 Down syndrome (trisomy 21)
 - 1.22 Edward syndrome and Patau syndrome (trisomy 18, trisomy 13)
 - 1.23 Other trisomies and partial trisomies of the autosomes, not elsewhere classified (includes pathogenic duplications, unbalanced translocations and insertions)
 - 1.24 Monosomies and deletions from the autosomes, not elsewhere classified (includes pathogenic deletions e.g. 22q11.2 deletion syndrome (diGeorge syndrome), Wolff-Hirschorn syndrome, Cri-du-chat syndrome
 - 1.25 Turner syndrome (monosomy X)
 - 1.26 Other sex chromosome abnormalities (e.g. Klinefelter syndrome)
 - 1.28 Other chromosomal abnormalities, not elsewhere specified (includes Fragile X syndrome, imprinting syndromes, triploidy)
 - 1.29 Unspecified
- 1.3 Genetic anomaly
 - 1.31 Genetic condition, specified (includes inborn errors of metabolism e.g. Tay-Sachs disease;)
 - 1.32 Syndrome/association with demonstrated chromosomal/gene anomaly
 - 1.39 Genetic condition, unspecified

Definitions and Rules:

This category includes deaths in which a major congenital anomaly, whether structural or chromosomal, is considered to have been the reason for the death. All categories correspond to the ICD10 numbering in Chapter XVII Congenital malformations, deformations and chromosomal abnormalities (Q00-Q99) as presented in ICD-PM¹¹





If termination of pregnancy was undertaken as a result of the anomaly include the digit "09" at the end the numerical classification e.g. Termination of Trisomy 21 (Down Syndrome) 1.2109. All terminations of pregnancy for congenital anomalies regardless of the causal link to perinatal death are also classified here. With mapping to ICD coding, non-lethal abnormalities may be identified.

Chromosomal and genetic testing are categorised separately, in recognition of advances in prenatal screening and testing. The scope of genetic testing is widening to include some conditions that may not manifest with structural anomalies in the prenatal period (e.g. Fragile X syndrome). If there is both a chromosomal/genetic and structural abnormality, code for the chromosome or genetic condition with the structural condition as an associated condition. Results of genetic testing of unknown significance are captured under associated conditions.

The chromosomal abnormality category excludes deaths where molecular karyotyping identifies an anomaly which is not thought to be causal. Findings of genetic testing of unknown significance (variations of uncertain or unknown significance, VUS) are classified as associated conditions. Where there is both a chromosomal and a structural abnormality, classify according to the chromosome abnormality with the structural abnormality as an associated condition.

Specific examples:

Down syndrome (Trisomy 21) is classified as a Chromosome abnormality (Down syndrome 1.21). If a cardiac anomaly is also present, this would be an associated condition (1.12 cardiovascular system).

Vater and *Vacterl* are 1.18 Congenital malformations affecting multiple systems, specified. For syndromes where DNA testing is available and has been confirmed for VATER or CHARGE association classify as genetic condition 1.31, specified.

Hydrops Fetalis: Antibody related hydrops (Immune Hydrops) e.g. Rhesus or Kell incompatibility and Bart's haemoglobinopathy (alpha thalassemia) is coded under as 1.17.

Non immune hydrops if due to chromosomal/genetic anomalies, classify under 1.2 and appropriate sub-classification, e.g. 1.25 Turner syndrome.

Idiopathic hydrops fetalis as 1.192 Other specific congenital anomaly, hydrops fetalis, idiopathic.

If the hydrops is secondary to underlying structural pathology e.g. congenital heart abnormality, neuromuscular disorders, skeletal dysplasia (achondrogenesis) or infection–classify in appropriate systems.

Hydrops associated with monochorionic twins classify under 6.1 category.

Multiple anomalies: Where the multiple anomalies are a part of a chromosomal anomaly found in the decedent, e.g. cleft lip and palate with heart defect as in velocardiofacial syndrome associated with 22q11 deletion, they should be classified under Category 1.2 but only if chromosome testing confirms deletion.

Anterior wall defects: Omphalocoele (exomphalos), gastroschisis, and congenital diaphragmatic hernia are now classified under musculoskeletal anomalies (Category 1.15), in line with ICD10-PM.

If omphalocoele is an isolated anomaly classify 1.15; if associated with multiple structural anomalies classify as 1.18; if associated with aneuploidy e.g. trisomy 18, classify as 1.22.





Acquired CNS anomalies: Infection-related abnormalities should be classified under Category 2, e.g. microcephaly/hydrocephaly secondary to CMV or toxoplasma infections should be classified as Category 2.21 and 2.3 respectively.

Congenital intracranial haemorrhage/injury may be classified as Category 7.5 Fetal antenatal intracranial injury.

Disruptions due to amniotic band disruption sequence may cause extensive asymmetric injury to the cranium and brain. It may also present as an encephaly or encephalocele. Classify under Category 7.5 *Fetal antenatal intracranial injury* but if due to alloimmune thrombocytopenia Code as 1.17 Haematological.

Neuromuscular disorders: Classify under 1.11. These are a complex group that may include primary muscle anomalies, CNS anomalies – both acquired and primary - and metabolic abnormalities. Some are syndromic with recognised recurrence risk. Associated anomalies may include pulmonary hypoplasia, hydrops and cleft palate. The cause of death may have been respiratory failure but the death should be classified as the underlying abnormality.

If the underlying aetiology is known classify accordingly – e.g. *Fetal antenatal intracranial injury* Category 7.5.

Unspecified Congenital Abnormalities: Category 1.19 Congenital anomaly, unspecified covers those cases where an abnormality was stated as the cause but where insufficient information was available to classify under other categories.



2 Perinatal infection

- 2.1 Bacterial
 - 2.11 Group B Streptococcus
 - 2.12 E coli
 - 2.13 Listeria monocytogenes
 - 2.14 Spirochaetal e.g. Syphilis
 - 2.18 Other bacterial
 - 2.19 Unspecified bacterial
- 2.2 Viral
 - 2.21 Cytomegalovirus
 - 2.22 Parvovirus
 - 2.23 Herpes simplex virus
 - 2.24 Rubella virus
 - 2.25 Zika virus
 - 2.28 Other viral
 - 2.29 Unspecified viral
- 2.3 Protozoal e.g. Toxoplasma
- 2.5 Fungal
- 2.8 Other specified organism
- 2.9 Other unspecified organism

Definitions and Rules

In order to qualify for this category, there must be evidence of fetal or neonatal infection as in Table 1. Determination of perinatal infection.

This category aims to identify all perinatal deaths due to infection as the primary cause including perinatal deaths with infection following spontaneous preterm labour or rupture of the membranes. Deaths in preterm infants following spontaneous rupture of the membranes or labour not fulfilling the definition of infection should be classified under Category 10 *Spontaneous Preterm*.

Category 2.8 Other specified organism includes deaths due to other identified organisms other than those in Categories 2.1 to 2.5. Category 2.9 Other unspecified organism includes cases where there is an obvious infection however the organism was either not identified or not specified.

Examples:

Classify here: Prelabour rupture of the membranes at term, with birth following 24 hours of membrane rupture, neonatal pneumonia identified within 48 hours of birth, subsequent neonatal death, group B Streptococcus identified on vaginal and placental cultures. Classify as subcategory 2.11 Group B Streptococcus and PSANZ-NDC subcategory 4.13.

Classify here: Spontaneous rupture of membranes preterm followed by spontaneous labour at 26 weeks and stillbirth. Membranes were ruptured for 12 hours prior to birth. Fetal pneumonia was detected at autopsy and growth of E Coli from the lungs. Placental pathology showed chorioamnionitis and funisitis. Classify 2.12 with an associated condition as Category 10.11 Spontaneous preterm, with chorioamnionitis on placental histopathology.





Classify here: Spontaneous rupture of the membranes at 24 weeks gestation. Clinical chorioamnionitis ensued after 6 days of membrane rupture. Induction of labour was undertaken resulting in birth of a liveborn infant. Birthweight was 650gms. Active resuscitation was unsuccessful. No autopsy or placental pathology was undertaken. Classify as Category 2.11 with an associated category of Spontaneous preterm Category 10.13 and PSANZ-NDC unspecified congenital infection 4.19 with an associated classification of NDC 2.2 Extreme prematurity – Unsuccessful resuscitation.

Classify here: Spontaneous rupture of membranes at 14 weeks, with severe chorioamnionitis at 22 weeks. Labour was induced (with a live baby) and the baby was born without signs of life, the birthweight was 350gms. Autopsy findings of *E.coli* growth from lung fluid. Placental histopathology showed chorioamnionitis and funisitis. Classify as Category 2.1209.

Do not classify here: Spontaneous rupture of membranes at 21 weeks, with spontaneous onset of labour and birth at 22 weeks gestation. Baby was born without signs of life with a birthweight was 450gms No autopsy was undertaken. Placental histopathology showed chorioamnionitis (no funisitis), no organism was grown. Classify as Category 10.11

Do not classify here: Neonatal death from late onset (≥48 hrs of age) Group B Streptococcal disease in a term infant. Classify under Category 12. Neonatal death with no obstetric antecedent factor and PSANZ-NDC as 4.4. The organism involved (GBS) may be classified as an associated condition under NDC associated factors using Category 2 sub classifications as a pragmatic way of collecting organisms in acquired infection. Alternately (and more appropriately), the organism should be included in a minimum dataset for all perinatal deaths



Death type	Criteria for Perinatal and Acquired Infection category
Fetal	Histological confirmation of inflammation in cord (funisitis) or fetus (pneumonitis or pneumonia) with or without microbiological evidence of infection or Convincing clinical evidence of primary maternal infection and
	2b. Positive culture of a pathogen from mother or placenta (specimen taken aseptically between amnion and chorion)
Neonatal	A. Congenital Early onset infection (within 48 hours of birth), defined as: 1. Clinical signs in neonate consistent with sepsis and 2. Haematological changes consistent with sepsis and one or more of the following: 3a. Positive culture of a pathogen (bacterial or viral) from the neonate or 3b. Pathological evidence at autopsy or 3c. Positive serology or 3d. Positive culture of a pathogen from the mother or the placenta or 3e. Pneumonia without specified bacterial or viral pathogens NB: Some congenital viral infections may have onset later than 48 hours after birth B. Acquired Onset of infection at 48 hours or later, with criteria as above, but excluding 3d

Table 1. Determination of perinatal infection



3 Hypertension

- 3.1 Chronic hypertension: essential
- 3.2 Chronic hypertension: secondary, e.g. renal disease
- 3.3 Chronic hypertension: unspecified
- 3.4 Gestational hypertension
- 3.5 Pre-eclampsia
- 3.6 Pre-eclampsia superimposed on chronic hypertension
- 3.9 Unspecified hypertension

Definitions

The classification of *Hypertension* follows that of the Society of Obstetric Medicine of Australia and New Zealand¹² with the exceptions that unspecified subcategories have been included. The definitions are as follows:

Hypertension is diagnosed when the systolic blood pressure is \geq 140 mm Hg and /or diastolic blood pressure (Korotkoff V) is \geq 90 mm Hg. These blood pressures should be confirmed by repeated readings over several hours in a clinic or day assessment unit or after rest in hospital.

Gestational hypertension is defined as hypertension arising in pregnancy after 20 weeks gestation without any other feature of the multisystem disorder pre-eclampsia and which resolves within 3 months postpartum.

Pre-eclampsia may be defined as hypertension arising after 20 weeks gestation and the onset after 20 weeks gestation of one or more of: proteinuria, renal insufficiency, liver disease, neurological problems, haematological disturbances, fetal growth restriction. The hypertension will have returned to normal within 3 months postpartum.

Rules

This category includes deaths where the hypertensive disorder is considered the factor initiating the chain of events leading to the death. If placental abruption complicates a hypertensive disorder, the death is classified here, as the abruption is attributed to the hypertensive disorder. Specific placental pathology can be coded as associated conditions (see PSANZ-SB&ND Associated conditions list page 34)

This category excludes the circumstance when the hypertension is secondary to an underlying systemic disorder, e.g. Diabetes, where this is severe and uncontrolled (in which case, classify as subcategory 5.2 *Diabetes*, under *Maternal Conditions*). However, if the systemic disorder such as diabetes or gestational diabetes is mild or well controlled, and the death appeared to be due to hypertension or its complications, classify in this category. This category also includes hypertension secondary to renal disease as this often presents first with hypertension.





4 Antepartum haemorrhage (APH)

- 4.1 Placental abruption
- 4.2 Placenta praevia
- 4.3 Vasa praevia
- 4.9 APH of undetermined origin

Definitions

Placental abruption: The diagnosis of placental abruption is made clinically. Confirmation by evaluation of the placenta after delivery is not essential for assigning the death to abruption. Clinically features are classically with vaginal bleeding (although the bleeding may be concealed), abdominal pain, uterine contractions and tenderness¹³.

Placenta praevia: Placenta praevia is defined as a placenta that lies wholly or partly within the lower uterine segment diagnosed on ultrasound¹³. With improved diagnosis and management stillbirth as a result of bleeding for placental praevia is now rare.

Vasa praevia: Vasa praevia is the presence of unsupported fetal vessels below the fetal presenting part, where the cord insertion is velamentous¹³. Classically, vaginal bleeding following amniotomy with subsequent fetal bradycardia suggests vasa praevia. The diagnosis of vasa praevia can be confirmed by Doppler and endovaginal ultrasound studies if aberrant vessels over the internal cervical os are suspected¹³.

APH of undetermined origin: This category is used where insufficient information is available on the reason for the bleeding. However, there is convincing clinical evidence that the stillbirth was as a result of the bleeding¹³.

Rules

This category includes all perinatal deaths where the primary factor leading to the death was an APH.

Convincing clinical signs of abruption alone is sufficient to assign the category of 4.1 Abruption. If abruption occurs as a complication of a hypertensive disorder, the death is attributed to the hypertensive disorder (Category 3) with Category 4.1 Placental abruption as an associated condition. Other placental pathology thought to be contributory may also be classified under associated conditions Category 9.

Examples:

Classify here: A women presents at 38 weeks' gestation with abdominal pain, tense abdomen and uterine contractions and a fetal death diagnosed. Placental macroscopic examination showed a large adhesive clot however placental histopathology was inconclusive. Classify as 4.1 Placental abruption.





5 Maternal Conditions

- 5.1 Termination of pregnancy for maternal psychosocial indications
- 5.2 Diabetes
 - 5.21 Gestational diabetes
 - 5.22 Pre-existing diabetes
- 5.3 Maternal injury
 - 5.31 Accidental
 - 5.32 Non-accidental
- 5.4 Maternal sepsis
- 5.5 Antiphospholipid syndrome
- 5.6 Obstetric cholestasis
- 5.8 Other specified maternal conditions
 - 5.81 Maternal suicide
 - 5.88 Other specified maternal medical or surgical conditions

Definitions and Rules

Category 5 includes perinatal deaths attributed to any medical or surgical condition in the mother, or to its complications or treatment, excluding conditions elsewhere classified i.e. APH, hypertension. The subcategory 5.1 excludes terminations of pregnancy undertaken for medical indication including congenital and other complications (e.g. prolonged preterm rupture of membranes (PPROM) with severe infection) where a pregnancy is terminated and the fetus is not expected to survive. In this scenario the death is classified under the specific condition including termination of pregnancy due to a congenital anomaly (classified under Congenital Anomaly, Category 1) and other conditions such as severe chorioamnionitis following preterm rupture of the membranes at 20 weeks (classify 10.1 Spontaneous preterm); adding the coding number "9" to identify termination as described under "General rules for applying the PSANZ PDC System" on page 4.

Renal disease is not included as a separate subcategory here, but under Hypertension, subcategory 3.2, as it usually presents first as hypertension. Maternal conditions should only be attributed here if there is a high probability that they were the cause of death, e.g. a well-documented history of lupus obstetric syndrome with a previous stillbirth. Maternal substance use or smoking may be classified as an associated condition if there is a significant history (including alcohol, cocaine, and marijuana) and where it is reasonable to assume that the fetal or neonatal death may be linked.

Examples:

Classify here: Fetal death as a result of severe uncontrolled Type I Diabetes with mild preeclampsia classify as subcategory 5.22 with an associated condition of Hypertension, Category 3.5.





6 Complications of multiple pregnancy

- 6.1 Monochorionic twins
 - 6.11 Twin to twin transfusion syndrome (TTTS)
 - 6.12 Selective fetal growth restriction (FGR) (i.e. affecting only one twin)
 - 6.13 Monoamniotic twins (including cord entanglement)
 - 6.18 Other
 - 6.19 Unknown or unspecified
- 6.2 Dichorionic twins
 - 6.21 Early fetal death in a multiple pregnancy (<20 weeks gestation)
 - 6.22 Selective fetal growth restriction (FGR)
 - 6.28 Other
 - 6.29 Unknown or unspecified
- 6. 3 Complications of higher order multiples (3 or more fetuses)
 - 6.31 Twin to twin transfusion syndrome (TTTS)
 - 6.32 Selective fetal growth restriction (FGR)
 - 6.33 Monoamniotic multiples (including cord entanglement)
 - 6.34 Early fetal death in a multiple pregnancy (<20 weeks gestation)
 - 6.38 Other
 - 6.39 Unknown or unspecified
- 6.4 Complications where chorionicity is unknown
- 6.8 Other
- 6.9 Unspecified

Rules

For 6.12, 6.22 and 6.32 read Explanatory Notes under Associated Condition, Section 15, Fetal Growth Restriction.

Where one of the twins (or multiples) is growth restricted as a result of twin to twin transfusion syndrome, classify as 6.11, 6.31 and not 6.12 or 6.32 respectively. Where one or more of the twins (or multiples) is growth restricted from a known underlying cause, classify elsewhere as appropriate, e.g. classify under Category 9 if there is placental disease in one of dichorionic twins.





7 Specific perinatal conditions

- 7.1 Fetomaternal haemorrhage
- 7.2 Antepartum cord or fetal vessel complications (excludes monochorionic twins or higher order multiples)
 - 7.21 Cord vessel haemorrhage
 - 7.22 Cord occlusion (True knot with evidence of occlusion or other)
 - 7.28 Other cord complications
 - 7.29 Unspecified cord complications
- 7.3 Uterine abnormalities
 - 7.31 Developmental anatomical abnormalities (e.g. bicornuate uterus)
 - 7.38 Other
 - 7.39 Unspecified
- 7.4 Alloimmune disease
 - 7.41 Rhesus isoimmunisation
 - 7.42 Other red cell antibody
 - 7.43 Alloimmune thrombocytopenia
 - 7.48 Other
 - 7.49 Unspecified
- 7.5 Fetal antenatal intracranial injury
 - 7.51 Subdural haematoma
 - 7.52 Fetal antenatal ischaemic brain injury
 - 7.53 Fetal antenatal haemorrhagic brain injury
- 7.6 Other specific perinatal conditions
 - 7.61 Complications of antenatal, diagnostic or therapeutic procedures:
 - 7.611 Complications of prenatal diagnostic procedures (e.g. amniocentesis, chorionic villus sampling,) (e.g. rupture of membranes after amniocentesis)
 - 7.612 Complications of fetal ultrasound guided needle interventions (e.g. FBS/fetal transfusion, thoracocentesis, vesicocentesis, fetal cardiac valvoplasty, division of amniotic bands, fetal skin biopsy, unipolar/bipolar diathermy, RFA procedures)
 - 7.613 Complications of fetal shunt interventions (e.g. pleuroamniotic shunt, vesicoamniotic shunt)
 - 7.614 Complications of minimally invasive fetoscopic interventions (e.g. fetoscopic laser surgery for TTTS, FETO for CDH, laser ablation of posterior urethral valves)
 - 7.615 Complications of open maternal fetal surgery (e.g. open maternal fetal surgery for spina bifida)
 - 7.618 Other
 - 7.62 Termination of pregnancy for suspected but unconfirmed congenital anomaly
 - 7.63 Amniotic band
 - 7.68 Other
- 7. 9 Unspecified





Definitions

Category 7.22 Cord occlusion: A cord knot is where the cord becomes tangled with itself (or another cord in a multiple pregnancy) such that the vessels of the cord may be compromised. To be considered significant there should be evidence of congestion or haemorrhage in the cord, and/or changes in the placenta such as fetal vessel thrombosis or villous oedema to suggest vascular compromise. A knot could cause death without these changes but not every knot causes fetal compromise and therefore should not be accepted as a cause of death without further evidence as above, or strong clinical suspicion by the delivering clinician based on CTG or other changes during delivery. Cord accidents usually only account for a few percent of perinatal deaths.

Other cord compression: For stillbirths, and also neonatal deaths as a result of hypoxic ischaemic encephalopathy (HIE), where the cord is found to be tightly around neck or body with skin blanching (indicating significant cord compression) classify as 7.28.

Category 7.21 includes cord haemorrhage following cordocentesis, umbilical cord ulceration leading to cord haemorrhage, and torn velamentous vessels.

Rules

This category includes deaths in which the specific perinatal condition present was thought to be the cause of death. The category excludes perinatal deaths with a major congenital anomaly. Cord complications during labour and other complications of twins e.g. head entrapment in labour should be categorised under *Hypoxic Peripartum Death*, subcategory 8.18.

Examples:

Do not classify here: Spontaneous prelabour rupture of membranes (ROM) at 33 weeks, with immediate cord prolapse and fetal death. Categorise as Spontaneous Preterm Category 10 as the cord complication occurred as a result of the preterm ROM. Cord prolapse is classified as an associated condition.





8 Hypoxic peripartum death

- 8.1 With intrapartum complications (sentinel events)
 - 8.11 Uterine rupture
 - 8.12 Cord prolapse
 - 8.13 Shoulder dystocia
 - 8.14 Complications of breech presentation
 - 8.15 Birth trauma
 - 8.16 Intrapartum haemorrhage
 - 8.18 Other
- 8.2 Evidence of significant fetal compromise (excluding other complications)
- 8.3 No intrapartum complications and no evidence of significant fetal compromise identified
- 8.9 Unspecified hypoxic peripartum death

Definitions and rules

This category includes both intrapartum fetal deaths and neonatal deaths as a result of acute or chronic hypoxia in babies without major congenital anomalies or other major conditions such as antepartum haemorrhage at a gestation in which survival in the context of the birth would be expected (typically of >28 weeks gestation or >1000g birthweight). If placental pathology is identified which resulted in fetal compromise and death then classify under the relevant category i.e. Category 9 Placental pathology or Category 4 Antepartum haemorrhage.

Where intrapartum fetal death or neonatal death occurs following preterm spontaneous onset of labour or rupture of membranes which fulfils the definition of Infection then classify under Category 2. If not fulfilling the criteria for infection and less than 24 weeks then classify under Category 10 Spontaneous preterm.

Neonatal deaths as a result of hypoxic ischaemic encephalopathy and otherwise unexplained severe cardiorespiratory depression at birth are included here. Where possible, evidence for intrapartum hypoxia should include fetal, umbilical artery or early neonatal (within one hour) blood gases showing evidence of a severe metabolic acidosis. Otherwise peripartum death might also be due to non-hypoxic causes, e.g. infection or chronic ischaemia but wrongly assumed to be due to acute hypoxia.

There may have been intrapartum complications (subcategory 8.1), or no intrapartum complications but with evidence of non-reassuring fetal status (subcategory 8.2), or no intrapartum complications or evidence of non-reassuring fetal status (subcategory 8.3). A specific major intrapartum complication, such as uterine rupture, cord prolapse or shoulder dystocia, is required for inclusion as subcategory 8.1. However, if there were no apparent intrapartum complications (as defined in category 8.1) but there was evidence of placental insufficiency antenatally, then the death should be attributed to Category 9. In this case Category 8 is captured as an associated condition.

If there is insufficient information about fetal wellbeing or intrapartum complications, classify as subcategory 8.9 *Unspecified hypoxic peripartum death*.

Evidence of non-reassuring fetal status is defined as abnormal fetal heart rate, fetal scalp pH/lactate, fetal pulse oximetry without intrapartum complications.





The term 'non-reassuring fetal status' has been used in preference to the term 'fetal distress' as 'clinical signs often poorly predict a compromised fetus and continued use of this latter term may encourage wrong assumptions or inappropriate management' 14,15.

Examples:

Classify here: No known problems prior to labour at gestation 38 weeks. Severe fetal heart rate decelerations in second stage of labour, without other major complication. Baby is born with no signs of life with a birthweight of 3500gm, placental histopathology did not identify any significant pathology and no autopsy was performed. Classify as subcategory 8.2.

Classify here: No known problems prior to labour at 36 weeks. No evidence of intrapartum fetal distress. At birth, the baby shows signs of severe respiratory depression and hypoxia. Subsequently develops encephalopathy and multiorgan failure and dies on Day 10 of life. Placental histopathology did not identify any significant pathology and no autopsy was performed. Classify as subcategory 8.3 and PSANZ NDC as 5.1.

Do not classify here: Spontaneous membrane rupture at 22 weeks' gestation, severe oligo hydramnios with positional deformities shown on ultrasound at 26 weeks. Labour and birth at 26 weeks gestation of a baby boy weighing 700gms and was not able to be resuscitated. Placental pathology showed chorioamnionitis but no organisms identified on placental culture or baby blood cultures. Classify as 10.11 Spontaneous preterm and PSANZ NDC as Category 2.2 Not resuscitated.

Do not classify here: No complications during pregnancy. Spontaneous preterm labour and birth at 38 weeks gestation. Intrapartum fetal distress in second stage and delivered by forceps. Baby boy weighing 2200gms, Apgars 1 and 4, mechanically ventilated and admitted to NICU. Seizures commenced at 2 hrs and active management ceased at 24 hrs due to poor prognosis. Placental pathology showed fetal vascular malperfusion and mild chorioamnionitis however no organisms were identified on culture of the placental or baby. Classify as 9.2 Placental dysfunction and PSANZ NDC 5.1 Hypoxic ischaemic encephalopathy/Perinatal asphyxia





9 Placental dysfunction or causative placental pathology

- 9.1 Maternal vascular malperfusion
- 9.2 Fetal vascular malperfusion
- 9.3 High grade villitis of unknown etiology (VUE)
- 9.4 Massive perivillous fibrin deposition/maternal floor infarction
- 9.5 Severe chronic intervillositis (Histiocytic intervillositis)
- 9.6 Placental hypoplasia (small-for gestation placenta)
- 9.7 No causal placental pathology demonstrated, with antenatal evidence of poor placental function identified (such as abnormal fetal umbilical artery Doppler)
- 9.8 Placental pathological examination was not performed, with antenatal evidence of poor placental function was identified (such as abnormal fetal umbilical artery Doppler)
- 9.9 Other placental pathology (e.g. multiple pathologies with evidence of loss of placental function leading to death)

Definitions

This category is based on the Amsterdam Placental Workshop Group Consensus Statement⁹.

Category 9.1 *Maternal vascular malperfusion (MVM)*. Placental features considered to be indicative of MVM include both gross and microscopic findings. Gross findings include placental hypoplasia, infarction, and retroplacental haemorrhage.

Any infarction seen in a preterm placenta and, at term, anything more than 5% of non-peripheral infarction should be classified as a cause. Although marginal infarcts in a term placenta may have less meaning than in a preterm placenta, they should be classified as an associated condition. *Microscopic findings* include abnormalities of villous development, which can be separated into distal villous hypoplasia, and accelerated villous maturation (vide infra), and infarcts. It should be recognized that many of these histologic findings will coexist in some placentas.

Category 9.6 Placental hypoplasia is reflected by a placental weight that is low for the stated gestational age and context (weight <10th centile) and/or a thin cord (<10th centile or <8-mm diameter at term).

Category 9.7 and 9.8 includes stillbirths or neonatal deaths where clinical evidence of poor placental function sufficient to explain the death was identified however significant causal pathology of the placental was not demonstrated or placental histopathology was not performed. Clinical evidence of poor placental function is defined as evidence of placental disease either on antenatal ultrasound studies or biochemistry. This former can include evidence of reduced maternal (uterine artery) or fetal (umbilical artery, ductus venosus, middle cerebral artery Doppler) vascular perfusion on Doppler studies. The latter can include angiogenesis-related factors such as s-Flt-1/PIGF; further clinical evaluations may clarify which biochemical markers robustly identify placental dysfunction.

Category 9.9 includes multiple pathologies with evidence of loss of placental function leading to death. It excludes pathologies listed in 9.1 to 9.8. Where one or more pathologies listed under 9.1-9.8 are identified, a single pathology must be classified as the primary cause of death with the additional pathologies classified as associated conditions (see Category 16 page 34).





Rules

This category includes perinatal deaths where placental dysfunction is considered the underlying cause of the death. It excludes perinatal deaths as a result of an identified maternal or fetal condition where the death is classified according to the condition (e.g. Pre-Eclampsia, Pre-existing hypertension). It should exclude pathology which is not thought to be causal, and also amniotic fluid infection/acute chorioamnionitis. Placental pathology which is thought to be contributory rather than causal should be classified as an associated condition (See Associated conditions page 34).

It is acknowledged that multiple pathologies may exist. In these circumstances a dominate pathology needs to be identified and classified as the main cause and others as associated conditions. This category overrides deaths following intrapartum related events as defined in Category 8 Hypoxic peripartum deaths.

Examples:

Classify here: Normal pregnancy. Spontaneous preterm labour and birth at 40 weeks gestation. Non-reassuring fetal status in second stage ensued and birth was by emergency caesarean section. Baby boy weighing 2600gms, Apgars 2 and 4, mechanically ventilated and admitted to NICU with subsequent diagnoses of meconium aspiration and persistent pulmonary hypertension of the newborn. Despite intensive care the baby died at 12 hrs of age. Placental pathology showed massive perivillous fibrin deposition/maternal floor infarction and mild chorioamnionitis, no organisms were identified on placental culture or baby blood cultures. Classify as 9.4 Massive perivillous fibrin deposition/maternal floor infarction and PSANZ NDC 3.3 Primary persistent pulmonary hypertension, with an Associated condition of Fetal growth restriction.

Do not classify here: Normal pregnancy until maternal presentation at 40 weeks' gestation with decreased fetal movements and abdominal pain. Antepartum fetal death was diagnosed and spontaneous labour ensued shortly after. A baby girl was born, mildly macerated, weighing 3400gms. Placental pathology showed massive abruption. Maternal investigations were normal. No organisms were identified on placental culture or baby blood cultures. Classify as APH Abruption 4.1.



10 Spontaneous preterm labour or rupture of membranes (<37 weeks gestation)

- 10.1 Spontaneous preterm
 - 10.11 With histological chorioamnionitis
 - 10.12 Without histological chorioamnionitis
 - 10.13 With clinical evidence of chorioamnionitis, no examination of placenta
 - 10.17 No clinical signs of chorioamnionitis, no examination of placenta
 - 10.19 Unspecified or not known whether placenta examined
- 10.2 Spontaneous preterm preceded by premature cervical shortening

Definitions

Clinical evidence of chorioamnionitis is defined as maternal fever (≥38°C) associated with one or more of the following symptoms or signs: maternal or fetal tachycardia, uterine tenderness, malodorous amniotic fluid, and maternal leukocytosis or raised C-reactive protein¹⁶⁻¹⁸.

The diagnosis of histological chorioamnionitis should only be made when there is histological evidence of inflammation or microbiological evidence of infection of the placenta and membranes.

The subcategory of premature cervical shortening is reserved for those circumstances where the primary event appears to be cervical change based on clinical or ultrasound findings. This may occur as consequence of pre-existing damage to the cervix from a surgical procedure, due to a congenital structural cervical anomaly (with or without uterine anomaly) or clinically determined from previous obstetric history and/or clinical factors in the current pregnancy.

Rules

Deaths of normally formed, appropriately grown preterm babies following spontaneous onset of preterm labour or spontaneous rupture of membranes, irrespective of induction of labour or mode of delivery (e.g. elective caesarean section). There should be no evidence of fetal or neonatal infection (see Table 1 Determination of perinatal infection), otherwise classify under Category 2 *Perinatal Infection*. Careful examination of the placenta macroscopically and microscopically is recommended.

In cases where there is histological evidence of chorioamnionitis with or without evidence of clinical chorioamnionitis, classify as subcategory 10.11. In cases of clinical chorioamnionitis where placental pathological examination was not performed or it is not known whether the placenta was examined, classify as subcategory 10.13.

Where cervical incompetence is followed by spontaneous preterm labour or ROM classify as 10.2 as opposed to 10.1. There may be some bleeding at the time of onset of labour, or earlier in pregnancy, but not in amounts to warrant the antecedent cause being attributed to *Antepartum Haemorrhage* Category 4. Early bleeding, which is often associated with preterm premature rupture of the membranes may be classified as an associated condition (see page 34).





Examples:

Classify here: Spontaneous labour at 26 weeks, no apparent explanation, and membranes intact. Vaginal delivery after 6 hours of membrane rupture, no evidence of intrapartum hypoxia or chorioamnionitis; subsequent early neonatal death from respiratory distress syndrome. Classify here as subcategory 10.12 Spontaneous preterm with intact membranes, or membrane rupture, without chorioamnionitis on placental histopathology and NDC: Category 3.1

Do not classify here: Spontaneous onset of labour at 28 weeks with intact membranes. No cause identified for preterm labour. Delivery following 24 hours of membrane rupture. Maternal intrapartum pyrexia. Chorioamnionitis and <u>funisitis</u> on placental histology, no organism identified. Classify as Category 2.9 Perinatal Infection; other unspecified organism

Do not classify here: Alive at the onset of spontaneous labour at 31 weeks, no apparent explanation, and membranes intact. After 12 hrs, continuous intrapartum fetal monitoring showed deep decelerations and emergency caesarean section undertaken. Baby girl weighing 1700g was stillborn and could not be resuscitated. Placental pathology showed chorioamnionitis (no funisitis) no organisms were identified, and no other pathology was demonstrated. No autopsy was performed. Macroscopic examination of the baby was normal, no maceration. Classify as Category 8.2 Hypoxic peripartum death; Evidence of significant fetal compromise (excluding other complications).



11 Unexplained antepartum fetal death

- 11.1 Unexplained antepartum fetal death despite full investigation
- 11.2 Unclassifiable antepartum fetal death with incomplete investigation
- 11.3 Unclassifiable antepartum fetal death due to unknown level of investigation

Rules

This category applies to fetal death prior to the onset of labour where no cause for the death was identified. Antepartum fetal death with associated placental pathology (i.e. not thought to be causative) are coded as associated conditions.

Category 11.1 Unknown antepartum fetal death despite full investigation.

An antepartum fetal death where no cause of death was identified following (as a minimum): comprehensive maternal and pregnancy history; histopathology of the placenta and cord; full autopsy; karyotype/cytogenetics; and testing for Feto-Maternal Haemorrhage (Kleihauer or flow cytometry).

Category 11.2 is used where none or some of the above investigations were performed and Category 11.3 is used where it is unknown/unclear if these investigations were performed or the results were unavailable.

Whether or not each of the above tests were performed should be recorded to identify areas of practice improvements and future research. The minimum dataset for perinatal deaths as defined in the Australian Perinatal Mortality Audit Tool APMAT (see Appendix E – Australian Perinatal Mortality Audit Tool) and the New Zealand PMMRC audit form 19 (Appendix E – Rapid reporting form for a perinatal death – baby and Appendix E – Rapid reporting form for a perinatal death – mother) includes these data fields.

Examples:

Classify here: Intrauterine Fetal Death (IUFD) at 37 weeks, with no maternal conditions, no antepartum haemorrhage, with membranes intact, before onset of labour, where no cause of death was identified following full investigation (comprehensive maternal and pregnancy history; histopathology of the placenta and cord; full autopsy; karyotype/cytogenetics; and Kleihauer) Classify as Unexplained Antepartum Fetal Death, subcategory 11.1.

Intrauterine Fetal Death (IUFD) at 40 weeks, with no maternal conditions, no antepartum haemorrhage, with membranes intact, before onset of labour, where no cause of death was identified and perinatal death investigations were incomplete (e.g. *No karyotype/cytogenetics*) Classify as Unexplained Antepartum Fetal Death, subcategory 11.2.

Do not classify here: Spontaneous ROM at 27 weeks, no significant maternal conditions present, subsequent IUFD prior to onset of labour. No chorioamnionitis on examination of the placenta. Classify as subcategory 10.12 Spontaneous preterm labour or ROM (<37 weeks gestation); without histological chorioamnionitis.





12 Neonatal death without obstetric antecedent

- 12.1 Neonatal death with no obstetric antecedent factors despite full investigation
- 12.2 Neonatal death unclassifiable as to obstetric antecedent with incomplete investigation
- 12.3 Neonatal death unclassifiable as to obstetric antecedent due to unknown level of investigation

Rules

This category includes neonates where no obstetric antecedent factors (according to the PDC list) were identified as contributing to the death.

Category 12.1 applies to a neonatal death where not obstetric antecedent factor was identified following negative findings for the following (as a minimum): comprehensive maternal and pregnancy history and full autopsy.

Category 12.2 is used where the full autopsy was not performed and Category 11.3 where it is unknown if they were performed or the results were unavailable.

NB: Whether a PDC code is assigned or not, all neonates require a neonatal cause of death according to the PSANZ NDC to be assigned. The NDC provides information on the causes and associated conditions present in the neonatal period.

Examples:

Classify here: Baby boy born at term weighing 3.5kg was discharged home well on Day 2 of life. On day 27, the baby was found dead in his cot by the parents and following full investigation was classified as SIDS. Please refer to the NDC to classify the neonatal cause of death.

Classify here: Baby boy born at 38 weeks weighing 3kg was discharged home well. On day 10, the baby became unwell and died. Blood cultures and CSF were positive for Group B Streptococcus. Please refer to the NDC to classify the neonatal cause of death and classify as 4.1.

Do not classify here: Neonatal death on Day 7 of a 29 week baby girl with severe fetal growth restriction and reverse end diastolic flow delivered by emergency caesarean section who developed fulminating necrotising enterocolitis. Placental pathology showed high grade villitis of unknown etiology (VUE). Classify as Category 9.3 with the PSANZ Associated condition of Fetal growth restriction and NDC Category 6.1 Necrotising enterocolitis.





2 PSANZ-NDC Classification including rules and definitions

The Neonatal Death Classification has been developed for use in conjunction with the PSANZ Perinatal Death Classification in order to identify the underlying and associated neonatal conditions as well as the underlying and associated maternal conditions for neonatal deaths. For example, a mother who has an antepartum haemorrhage at 32 weeks gestation delivers a 1500g infant who thrives in the neonatal nursery but subsequently acquires a lethal nosocomial infection: the obstetric antecedent is antepartum haemorrhage, but neonatal death classification is subcategory 4.4 Acquired Bacteria. Both APH and neonatal nosocomial infection are important conditions on which to focus prevention strategies.

1 Congenital anomaly (please refer to PDC)

2 Periviable infants (typically <24 weeks)</p>

- 2.1 Not resuscitated (including infants where there is an antenatal plan for no resuscitation at birth)
- 2.2 Unsuccessful resuscitation
- 2.9 Unspecified or not known whether resuscitation attempted

This group includes infants deemed too immature or too small for resuscitation or continued life support beyond the delivery room. Resuscitation in this context means the use of positive pressure ventilation.

3 Cardio-respiratory disorders

- 3.1 Hyaline membrane disease / Respiratory distress syndrome (RDS)
- 3.2 Meconium aspiration syndrome
- 3.3 Primary persistent pulmonary hypertension
- 3.4 Pulmonary hypoplasia
- 3.5 Pulmonary haemorrhage
- 3.6 Air leak syndromes
 - 3.61 Pneumothorax
 - 3.62 Pulmonary interstitial emphysema
 - 3.68 Other
- 3.7 Patent ductus arteriosus
- 3.8 Chronic neonatal lung disease (typically, bronchopulmonary dysplasia)
- 3.9 Other
 - 3.91 Neonatal anaemia/hypovolaemia

Definitions and Rules

Subcategory 3.1 Hyaline membrane disease / Respiratory Distress Syndrome (RDS) is used for deaths of infants who were receiving mechanical ventilation for acute RDS at the time of death or at the time of the complication such as pulmonary haemorrhage, sepsis, pneumothorax or necrotizing enterocolitis.

Neonates with resolving RDS, i.e. who are past the acute phase of the disease and are stable or improving, but who are still on low rate ventilation for immature lungs, extreme prematurity or apnoea, or who no longer require mechanical ventilation, and who developed a complication which led to the death should be classified according to that particular complication.





3.4 Pulmonary hypoplasia; this category includes pulmonary hypoplasia secondary to preterm prolonged rupture of the membranes. Congenital pulmonary airway malformation (CPAM), formerly known as congenital cystic adenomatoid malformation of the lung (CCAM) would be classified as 1.16. Congenital diaphragmatic hernia is classified as 1.151

Categorisation as chronic neonatal lung disease (subcategory 3.5) should be reserved for infants with deteriorating lung function and major chest X-ray changes consistent with bronchopulmonary dysplasia.

Examples:

Classify here: A 26-week gestation infant with RDS receives mechanical ventilation (SIPPV R50, P20/5, Fi02 0.4), develops complications of pneumothorax requiring drainage, followed by a patent ductus arteriosus and dies on day 2 of life is classified as Category 3.1 with associated conditions classified as 3.61 *Pneumothorax* and 3.7 *Patent ductus arteriosus*.

Do not classify here: A 26 week gestation infant with RDS weaning off mechanical ventilator has a Grade IV Intraventricular Haemorrhage (IVH) with ventricular dilation on ultrasound on Day 5. She is successfully weaned to CPAP on Day 7 but requires re-ventilation for sepsis on Day 10 and on Day 21 has developing BPD and post hemorrhagic hydrocephalus (PHH) following which ventilation is withdrawn. Classification is dependent on the major reason for withdrawal of support. In this case 5.3 Post haemorrhagic hydrocephalus with an associated classification of 3.8 Chronic neonatal lung disease and 4.49 Sepsis.



4 Neonatal infection

- 4.1 Congenital/Perinatal bacterial infection (early onset<48 hrs)
 - 4.11 Blood stream infection/septicaemia
 - 4.111 Positive culture of a pathogen
 - 4.112 Clinical signs of sepsis + ancillary evidence but culture negative
 - 4.12 Bacterial meningitis
 - 4.13 Bacterial pneumonia
 - 4.15 Multiple site bacterial infection
 - 4.18 Other congenital bacterial infection e.g. gastroenteritis, osteomyelitis, cerebral abscess
 - 4.19 Unspecified congenital infection
- 4.2 Congenital/Perinatal viral infection
- 4.3 Congenital fungal, protozoan, parasitic infection
- 4.4 Acquired bacterial infection (late onset>48hrs)
 - 4.41 Blood stream infection/septicaemia
 - 4.411 Positive culture of a pathogen
 - 4.412 Clinical signs of sepsis + ancillary evidence but culture negative
 - 4.42 Bacterial meningitis
 - 4.43 Bacterial pneumonia
 - 4.48 Other acquired bacterial infection e.g. gastroenteritis, osteomyelitis
 - 4.49 Unspecified acquired infection
- 4.5 Acquired viral infection
- 4.6 Acquired fungal, protozoan, parasitic infection

Rules

This category is intended to be used in conjunction with the PDC Category 2 *Perinatal Infection* to identify the organism causing the infection resulting in the death (including for an acquired infection). To take a pragmatic approach to storage of these data within the current system structure, in the case of a neonatal death from infection, the relevant NDC code can be stored as the primary neonatal condition and the PDC Category 2 code as an associated condition.





Determination of congenital and acquired neonatal infection

A. Congenital

Early onset infection (within 48 hours of birth), defined as:

1. Clinical signs in neonate consistent with sepsis

and

2. Haematological changes consistent with sepsis and one or more of the following:

3a. Positive culture of a pathogen (bacterial or viral) from the neonate

or

3b. Pathological evidence at autopsy

or

3c. Positive serology

or

3d. Positive culture of a pathogen from the mother or the placenta. Swap taken aseptically between amnion and chorion.

or

3e. Pneumonia without specified bacterial or viral pathogens

NB: Some congenital viral infections may have onset later than 48 hours after birth.

B. Acquired/nosocomial

Onset of infection at 48 hours or later, with criteria as above, but excluding 3d.

Table 4. Determination of infection



5 Neurological

- 5.1 Hypoxic ischaemic encephalopathy/Perinatal asphyxia
- 5.2 Cranial haemorrhage
 - 5.21 Intraventricular haemorrhage
 - 5.22 Subgaleal haemorrhage
 - 5.23 Subarachnoid haemorrhage
 - 5.24 Subdural haemorrhage
 - 5.28 Other intracranial haemorrhage
- 5.3 Post haemorrhagic hydrocephalus
- 5.4 Periventricular leukomalacia
- 5.8 Other

Definitions and Rules:

Hypoxic ischaemic encephalopathy/Perinatal asphyxia:

Inclusion as hypoxic ischaemic encephalopathy or perinatal asphyxia usually requires a defining asphyxial event +/- evidence of severe non-reassuring fetal status and encephalopathy.

Examples of defining asphyxial events:

Massive antepartum haemorrhage from abruption (4.1), placenta praevia (4.2) or ruptured vasa praevia (4.3), breech presentation (8.14) or delivery with complications, e.g. cervical constriction ring or difficult delivery, feto-maternal haemorrhage (7.1), twin-twin transfusion (6.11).

Where possible, evidence for perinatal asphyxia should include fetal, umbilical artery or early neonatal blood gases (within one hour) showing evidence of a severe metabolic acidosis. Otherwise peripartum death might also be due to non-hypoxic causes, e.g. infection or chronic ischaemia but wrongly assumed to be due to acute hypoxia. In the absence of a defining asphyxial event every effort must be undertaken to exclude alternative diagnosis.





6 Gastrointestinal

- 6.1 Necrotising enterocolitis (NEC)
- 6.2 Short gut syndrome
- 6.3 Gastric or intestinal perforation (excluding NEC)
- 6.4 Gastrointestinal haemorrhage
- 6.8 Other

Definitions and Rules

When Short gut syndrome is a consequence of NEC or gastroschisis (1.152) then classify as Category 6.2 Short gut syndrome for the cause and other conditions as associated. Short gut syndrome Category 6.2 includes major intestinal infarction (such as midgut volvulus (1.14).





7 Other

- 7.1 Sudden unexpected death in infancy (SUDI)
 - 7.11 Sudden Infant Death Syndrome (SIDS)
 - 7.112 SIDS Category IA: Classic features of SIDS present and completely documented
 - 7.113 SIDS Category IB: Classic features of SIDS present but incompletely documented
 - 7.114 SIDS Category II: Infant deaths that meet category I except for one or more features
 - 7.12 Unclassified Sudden Infant Death in the neonatal period
 - 7.121 Bed sharing/unsafe sleep
 - 7.122 Not bed sharing
 - 7.19 Unknown/Undetermined
- 7.2 Multisystem failure
 - 7.21 Secondary to intrauterine growth restriction
 - 7.28 Other specified
 - 7.29 Unspecified/undetermined primary cause or trigger event
- 7.3 Trauma
 - 7.31 Accidental
 - 7.32 Non accidental
 - 7.39 Unspecified
- 7.4 Treatment complications
 - 7.41 Surgical
 - 7.42 Medical
- 7.5 Unsuccessful resuscitation in infants of 28 weeks gestation or more without an obvious sentinel event
- 7.8 Other specified

Definitions

7.1 SIDS and 7.91 Unclassified Sudden Infant Death are defined according to the new SIDS classification system by Krous et al²⁰.

General Definition of SIDS

SIDS is defined as the sudden unexpected death of an infant <1 year of age, with onset of the fatal episode apparently occurring during sleep, that remains unexplained after a thorough investigation, including performance of a complete autopsy and review of the circumstances of death and the clinical history.

Category IA SIDS: Classic Features of SIDS Present and Completely Documented

Category IA includes infant deaths that meet the requirements of the general definition and also all of the following requirements.

Clinical:

- More than 21 days and <9 months of age;
- Normal clinical history, including term pregnancy(gestational age of \geq 37 weeks);
- Normal growth and development.





 No similar deaths among siblings, close genetic relatives (uncles, aunts or first-degree cousins), or other infants in the custody of the same caregiver.

Circumstances of Death:

- Investigation of the various scenes where incidents leading to death might have occurred and it is determined that they do not provide an explanation for the death.
- Found in a safe sleeping environment, with no evidence of accidental death.

Autopsy:

- There is an absence of potentially fatal pathologic findings. Minor respiratory system
 inflammatory infiltrates are acceptable; intrathoracic petechial haemorrhage is a
 supportive but not obligatory or diagnostic finding.
- There is no evidence of unexplained trauma, abuse, neglect, or unintentional injury.
- There is no evidence of substantial thymic stress effect (thymic weight of <15g and/or moderate/severe cortical lymphocyte depletion). Occasional "starry sky" macrophages or minor cortical depletion is acceptable.
- Results of toxicologic, microbiologic, radiologic, vitreous chemistry, and metabolic screening studies are negative.

Category IB SIDS: Classic Features of SIDS Present but Incompletely Documented

Category IB includes infant deaths that meet the requirements of the general definition and also meet all of the criteria for category IA except that investigation of the various scenes where incidents leading to death might have occurred was not performed and or ≥ 1 of the following analyses was not performed: toxicologic, microbiologic, radiologic, vitreous chemistry, or metabolic screening studies.

Category II SIDS

Category II includes infant deaths that meet category I criteria except for ≥ 1 of the following.

Clinical:

- Age range outside that of category 1A or 1B (i.e., 0-21 days or 270 days [9 months] through first birthday);
- Similar deaths among siblings, close relatives, or other infants in the custody of the same caregiver that are not considered suspect for infanticide or recognised genetic disorders;
- Neonatal or perinatal conditions (for example, those resulting from preterm birth) that have resolved by the time of death.

Circumstances of Death:

Mechanical asphyxia or suffocation caused by overlaying not determined with certainty.

Autopsy:

- Abnormal growth and development not thought to have contributed to death;
- Marked inflammatory changes or abnormalities not sufficient to be unequivocal causes of death.

Unclassified Sudden Infant Death





The unclassified category includes deaths that do not meet the criteria for category I or II SIDS but for which alternative diagnoses of natural or unnatural conditions are equivocal, including cases for which autopsies were not performed.

Post-resuscitation cases

Infants found in extremis who are resuscitated and later die ("temporarily interrupted SIDS") may be included in the aforementioned categories, depending on the fulfilment of relevant criteria.

Rules

Subcategory 7.92 Other Unknown/Undetermined has been included to identify unknown causes of death which do not fulfil the criteria of Category 7.91. Subcategory 7.4 Other accident, poisoning or violence (postnatal) excludes cases of antepartum deaths which should be classified in Category 5 Maternal Conditions under subcategory 5.3 Maternal injury. Subcategory 7.8 Other specified is used to classify other identified conditions which are not included in subcategories 7.1 to 7.4.



7.8 PSANZ Associated Conditions

Following classification of the underlying cause of death according to the PSANZ-PDC for stillbirths and neonatal deaths, and in addition a PSANZ NDC for neonatal deaths, associated conditions thought to be contributory (but not causal) to the death should be classified. The associated conditions list includes the PSANZ-PDC categories and, in addition for neonatal deaths, the PSANZ-NDC categories and other conditions which may be contributory to stillbirth as listed below in Categories 13-16.

Associated conditions for both stillbirths and neonatal deaths

Categories 1 -11 PSANZ PDC Plus the following additional categories:

13 Genetic testing results not diagnostic

- 13.1 Copy number variant of unknown or uncertain significance
- 13.2 No mutation identified matching phenotype
- 13.3 Tested for genetic mutations but failed
- 13.4 Not tested or not known if tested for genetic mutations

Explanatory/clarifying notes:

Where a pathogenic or a likely pathogenic mutation has been identified, this would have been classified under Category 1.2 Chromosomal anomaly as stated in the Definitions and Rules section of Category 1 Congenital anomaly. 1.31 and 1.34 are self-explanatory. 13.3 tested for genetic mutations but failed, refer to those tests that may have failed due to culture failure (with conventional cytogenetics) or poor DNA (with molecular techniques)

14 Associated placental pathology

- 14.1 Delayed villous maturation
- 14.2 Large chorioangioma
- 14.3 Early bleeding often leading to preterm prelabour ROM
- 14.8 Other associated placental pathology

Explanatory/clarifying notes:

Early bleeding is defined as bleeding in the second trimester (often on one or more occasions) which does not immediately lead to spontaneous birth or rupture of membranes.

15 Associated cord pathology

- 15.1 True knot (excluding histological evidence of causation)
- 15.2 Hypercoiled cord
- 15.3 Tethered cord
- 15.4 Velamentous insertion
- 15.8 Other associated cord pathology

16 Fetal Growth Restriction (FGR)

- 16.1 Autopsy evidence (brain:liver ratio equal to or greater than 4:1)
- 16.2 Antenatal ultrasound evidence of FGR
- 16.3 Clinical examination of the baby (by paediatrician, pathologist)
- 16.4 Birthweight (less than 10th centile for gestational age)
 - 16.41 Customised centiles²¹
 - 16.42 Population centiles^{22,23}

Explanatory/clarifying notes:

Fetal growth restriction is defined as:

- A brain:liver ratio equal to or greater than 4:1 at autopsy AND/OR
- Where antenatal ultrasound assessment has shown evidence of FGR (e.g. reduced growth velocity on serial biometry and/or abnormal utero-placental blood flow on Doppler ultrasound and reduced amniotic fluid volume) AND/OR
- 2. Clinical examination of the baby (by paediatrician, pathologist) AND/OR
- 3. Birthweight <10th centile for gestational age for livebirths or non-macerated stillbirths

Classifying FGR in stillbirths

It is also recommended that for fetal deaths, where possible, the gestational age on the date of death and not date of birth be used to define the presence of FGR.

For macerated stillbirths, in the absence of prior ultrasound evidence of FGR and where no autopsy has been performed or the brain:liver ratio is less than 4:1, the death should be classified as Unexplained Antepartum Death (Category 11), as the weight discrepancy may be a post mortem effect.

Customised or non-customised centiles

Either customised or non-customised centiles charts can be used to classify FGR as an associated condition under 16.2.1 or 16.2.2 respectively. Customised birthweight (CBW) centiles are being increasingly used to determine the presence of FGR²¹. However controversy around the use of customised centiles continues^{24,25} including concerns that customisation may mask pathology^{24,26}. It is recommended that the variables required for calculation of CBW (maternal age, ethnicity, height, weight, and fetal gestation and gender) be routinely collected to enable evaluation of birthweight according to CBW centiles. The recommended Australia population standards are those published by Dobbins et al²² and for preterm birth by Fenton et al²³.

17 Maternal risk factors (optional category)

- 17.1 Smoking
- 17.2 Substance use
- 17.3 High BMI
- 17.4 Maternal mental health disorder
- 17.5 Socioeconomic deprivation
- 17.6 Refugee or asylum seeker





Ideally risk factors would be included as part of a minimum dataset for all livebirths and stillbirths to enable ongoing assessment of the contribution of these factors to perinatal deaths. Further, inclusion of the PSANZ classification in this dataset for each perinatal death will provide a rich source of information for understanding causal pathways for maternal risk factors.

Associated conditions for neonatal deaths only

NDC Categories 1-6

In addition to the above for associated maternal/fetal conditions the NDC Categories 1- 6 can be used to assign associated neonatal conditions



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7.12 Appendices

Appendix A – Stillbirth investigations algorithm

Appendix B – Estimation of severity of feto-maternal haemorrhage

Appendix C – Placental examination; Accoucheur flow chart

Appendix D – Clinical examination of baby checklist

Appendix E – Australian perinatal mortality audit tool

Appendix F – New Zealand Rapid reporting form for a perinatal death - baby

Appendix G – New Zealand Rapid reporting form for a perinatal death - mother

Appendix H – Instructions on taking clinical photographs

Appendix I – Autopsy clinical summary form

Appendix J – Perinatal mortality classifications: Quick reference sheet

Appendix K – WHO mortality audit meeting code of practice declaration

Appendix L - Birthweight percentiles

Appendix M – Infant autopsy consent brochure

Appendix N – Information for health professionals seeking consent

Appendix O – RCPath Guidelines for Autopsy Investigation of Fetal and Perinatal Death

Appendix P - Placental histopathology reporting form

Appendix Q – Suspected genetic metabolic disorders

Appendix R – Screening for genetic metabolic disorders

Appendix S – Components of the genetic autopsy for investigations of metabolic disorders

Appendix T – Australian and New Zealand definitions of perinatal mortality

Appendix U – Changes on this version of the classifications

Appendix V – Development of PSANZ Perinatal Death Classification and PSANZ Neonatal Death Classification

Appendix W – Methods

Appendix X – Glossary of terms and abbreviations

Appendix Y – Contact details and regional coordinators 2017



