



Giornata Regionale SIDS, SIUD & ALTE



S.I.U.D. E S.I.D.S.

INQUADRAMENTO CLASSIFICATIVO E METODOLOGIA DI STUDIO

Dott. Vincenzo Nardini
U.O. Anatomia Patologica 2
Azienda Ospedaliero-Universitaria Pisana

LA SPEZIA, 27 NOVEMBRE 2010

Stillbirth and S.I.D.S.

Definitions relating to death in early life differ in different countries and in different jurisdictions.

It is important to be aware of them when comparing statistics or papers from different countries.

In order to identify ways in which perinatal deaths may be prevented, one needs a valid system of classification.

Stillbirth and S.I.D.S.

Definitions

Fetal death	< 22 week of gestation (WHO) < 24 w.g. (UK)	When the conceptus exhibits no signs of life after complete separation from the mother
Stillbirth	after 22 w.g. (WHO) after 24 w.g. (UK) early stillbirth < 28 w.g. late stillbirth > 28 w.g.	As above
Neonatal death	Death following live birth < 28 days (UK) < 30 days (USA)	Exhibition of signs of life after complete separation from the mother

According to different classifications, stillbirth is about ten times more common than sudden infant death syndrome (stillbirth rate in developed countries is estimated between 4.2 and 6.8 / 1000 births).

Although rates of sudden infant death syndrome have greatly fallen over the past 10-15 years, there has been a recent slight rise in the rate of stillbirth (S.I.U.D).

The aim of this presentation is to provide an overview of the classification (and the causes) of stillbirth and to try to point out possible maternal risk factors.

Classifications

Aberdeen clinico-pathological classification

- 1) Premature, cause unknown
- 2) Mature, cause unknown
- 3) Mechanical
- 4) Toxaemia
- 5) Antepartum haemorrhage
- 6) Maternal disease
- 7) Deformity (malformations)
- 8) Other causes

British Perinatal Mortality Survey

- 1) Congenital malformations
- 2) Blood group incompatability
- 3) Asphixyia (antepartum)
- 4) Asphixyia (intrapartum)
- 5) Intraventricular haemorrhage
- 6) Hyaline membranes
- 7) Massive pulmonary haemorr.
- 8) Pneumonia
- 9) Extrapulmonary infections
- 10) No anatomical lesions found

Baird D. et al. J Obset Gynaecol Br Emp 61: 433-448. 1954

Claireaux AE. Butterworths, London. Pp 191-211. 1962

Classifications

Naeye RL. JAMA 238: 228-229. 1977

Placental and Fetal P	athology Classification
1) Acute amniotic fluid infection syndrome	11) Rhesus erytroblastosis
2) Abruptio placentae	12) Birth trauma
3) Premature rupture of membranes	13) Polyhydramnios
4) Congenital anomalies	14) Caesarean section
5) Large placental infarct	15) Marginal sinus rupture
6) Intervillous thrombi of placenta	16) Severe fetal undernutrition
7) Umbilical cord compression	17) Uterine rupture
8) Cord knots	18) Posmaturity
9) Placental growth retardation	19) Congenital syphilis
10) Placenta praevia	20) Other disorders

Classifications

Wigglesworth classification

- 1) Macerated
- 2) Malformation
- 3) Immaturity
- 4) Asphixyia
- 5) Other

Wigglesworth J. Lancet II: 684-686. 1980

Extended Wigglesworth classification

- 1) Congenital defects
- 2) Unexplained antepartum fetal death 45,5 %
- 3) Intrapartum asphixyia, anoxia, trauma
- 4) Immaturity
- 5) Infection
- 6) Other specific causes
- 7) Unclassifiable

1,9 %

8) Problematic

Classifications

	 an-		000	MAN
		-		

- 1) Trauma
- 2) Infection
- 3) Placenta/Cord disease
- 4) Maternal immune system disease
- 5) Congenital malformations
- 6) Prematurity/Immaturity
- 7) Unclassifiable 18,2 %

ReCoDe

- A) Fetus
- B) Umbilical cord
- C) Placenta
- D) Amniotic fluid
- E) Uterus
- F) Mother
- G) Intrapartum
- H) Trauma
- I) Unclassified 14,3 %

Tulip

- 1) Congenital anomaly
- 2) Placenta
- 3) Prematurity/Immaturity
- 4) Infection
- 5) Other
- 6) Unknown **16,2 %**

De Galaan Roosen AEM et al. Eur J Obstet Gynaecol. 103: 30-6 2002 Gardosi J et al. BMJ 331: 1113-7 2005

Korteweg FJ. BJOG. 113: 393-401 2006

Risk factors for unexplained fetal death

1967-1976 1977-1998

Unexplained fetal mortality 0.24 % 0.16 %

All fetal deaths 30 % 40 %

Unexplained fetal death occurred later in gestation than explained and was associated with:

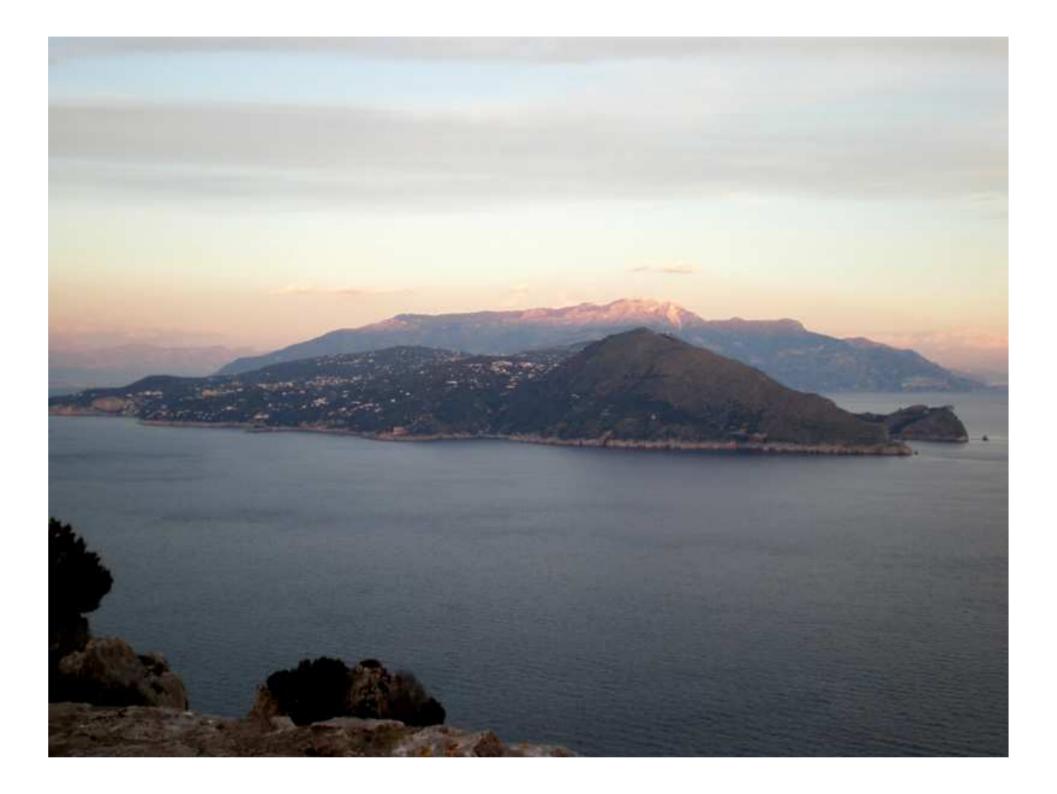
high maternal age;

multiparity;

low education (less than 10 yrs education against \geq 13 yrs);

low and high birth weight percentile

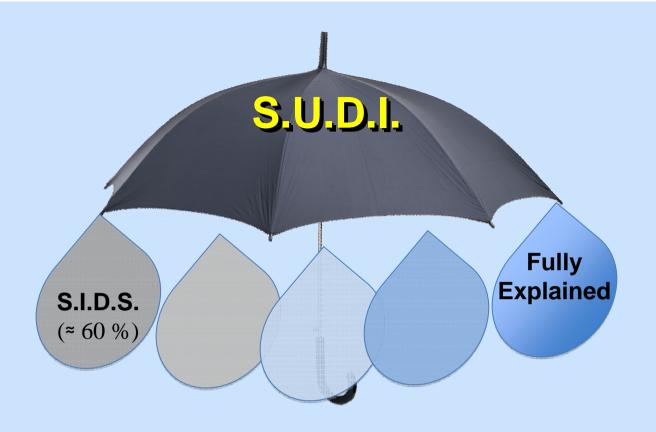
Rasmussen S. Early Hum Develop. 71: 39-52. 2003



S.U.D.I. (Sudden Unexpected Death in Infancy)

SUDI is a term used for all unexpected deaths, whether the explanation is immediate, determinable after a thorough investigation or remains unknown.

Traditionally all unexplained SUDI death have been classified as S.I.D.S., but more recently if certain risk factors are present (cosleeping, prone positioning, etc.) are preferred terms like: unascertained (UK), undetermined (Australia), unknown/unspecified (U.S.)



Sudden and Unexpected terms mean that infants have generally been either completely well or have been suffering from only an apparent minor illness before death occurred.

Infant deaths classified as S.U.D.I.		
Explained deaths	Rapid INFECTIONS	Meningitis, septicaemia, myocarditis, bronchopneumonia, massive gastro-enteritis, etc.
	Unrecognised pre-existing medical conditions	Metabolic disorders, unrecognised congenital abnormality, intestinal obstruction
	Accidental deaths	Road traffic accident, drowning, overlaying
	Non-Accidental deaths	Infanticide, neglect, abuse, etc
Unexplained deaths	S.I.D.S./unascertained	

S.I.D.S. (Sudden Infant Death Syndrome)

In 1970's was introduced the "three hit model" for SIDS suggesting that sudden and unexpected death may occur if three conditions are fulfilled simultaneously:

- 1. An infant is at a vulnerable development stage;
- 2. A predisposing endogenous factor(s) is present;
- 3. An exogenous trigger initiates the lethal process.

S.I.D.S. (Sudden Infant Death Syndrome)

This hypothesis was later modified including "genetic risk factors" or "genetic make up" as predisposing factors, investigating the significance of mtDNA changes, complement component C4 polymorphism, polymorphisms of the Il-10 gene, ...

Sudden deaths in early life (fetal, perinatal period, infancy and early childhood) may be the outcome of a variety of natural diseases as well as accidents and homicide.

In any sudden dependent D./S.I.D.S.), it is most important that a full evestigations is carried out and that their results are clearly presented.

S.I.D.S. (Sudden Infant Death Syndrome)

Definition

A SUDDEN AND UNEXPECTED DEATH OF AN INFANT

- > under one year of age;
- > the onset of the fatal episode was apparently associated with sleep;
- ➤ a thorough investigation of the case (including the performance of a complete autopsy, review of the circumstances of death and clinical history)

DOES NOT PROVIDE AN EXPLANATION FOR THE DEATH

Radiology is mandatory. It should be performed prior to the autopsy: antero-posterior projections and lateral views; (additional projections if necessary).

Necropsy examination should not be contemplated in establishments where radiography is not available.

From the literature, it is known that it contributes to the diagnosis in about 3-4 % of infant deaths and in about 25 % of unnatural deaths

External examination

- weights and measurements;
- detailed external examination by region (injuries, changes of uncertain aetiology, petechiae, secretions emanating from the nose and mouth);

Photography High quality photographs are an important part of the record of any necropsy on fetus, infants or children and are particularly important when investigating sudden deaths.

Whole-body views give a useful visual reminder of body proportions and state of nutrition.

Microbiological investigations are an important part of the examination of SUDI (as early as possible)

Microbiological samples usefully taken in sudden death investigation

Bacteriological samples	Virological samples
Nose/throat/bronchial swabs Blood culture	Nose swabs/nasopharingeal aspirates Tracheal ring
Cerebro-spinal fluid	Lung
Lung	Heart
Large bowel or rectal swab	Small intestine
•••••	•••••

Toxicological investigations are not taken routinely in many jurisdictions as part of the investigation of SUDI.

They are more likely to be done when concerns are expressed in a police report.

	Useful toxicological samples in the investigation of SUDI	
Samples		Toxicological agent
Blood		Alcool; Common sedatives
Urine		Common analgesics; Recreational drugs
Lung		Volatiles
Hairs		Long-term drug ingestion
Stomacl	h contents	Common sedatives/analgesics

Biochemical and Metabolic investigations:

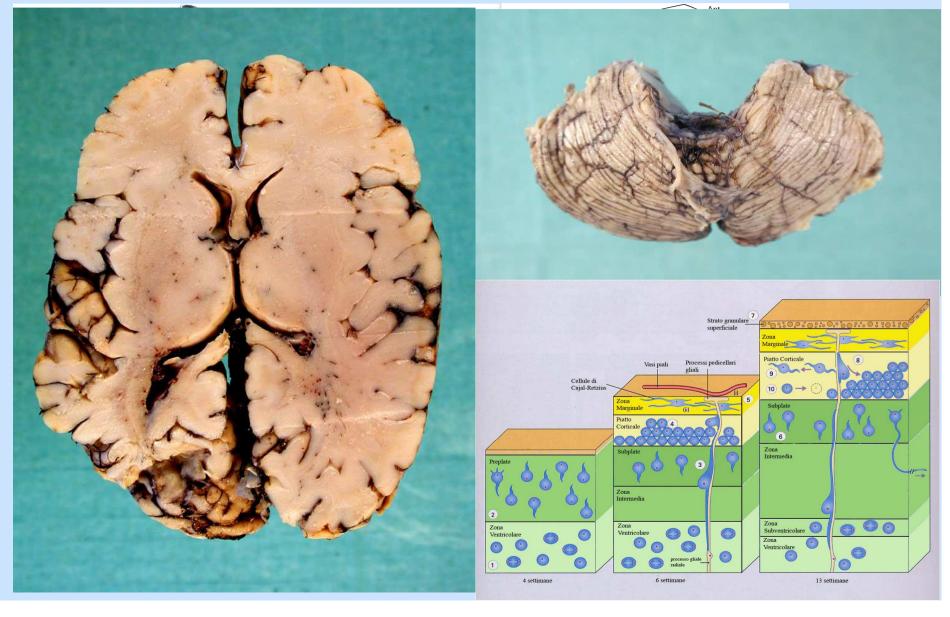
blood samples are unsatisfactory;

vitreous, urine and cerebrospinal fluid are much more usueful (often bladder may be empty in SUDI cases).

Dissection

The task is easier when a complete equipment is available (digital balance, complete forceps and scissors sets).

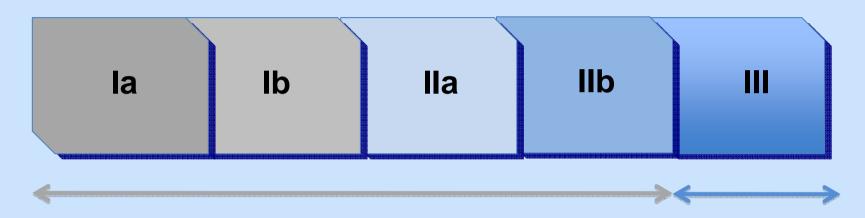
Relationship of organs are observed before dissection commences (some abnormal relationships may suggest a syndrome diagnosis).



The starting point for the classification of S.U.D.I. will be based on the *Avon clinico-pathological system*.

This System simply grades the unexplained S.U.D.I. from those deaths that are completely inexplicable (Ia) to those where a complete explanation has been found (III).

Avon clinico-pathological system.



Unexplained S.I.D.S.

Explained S.U.D.I.

Ia: No notable factors identified

Ib: Notable factors identified but not likely to have contributed to the death

IIa: Factor(s) identified that possibly contributed to the death

IIb: Factor(s) identified that probably contributed to the death

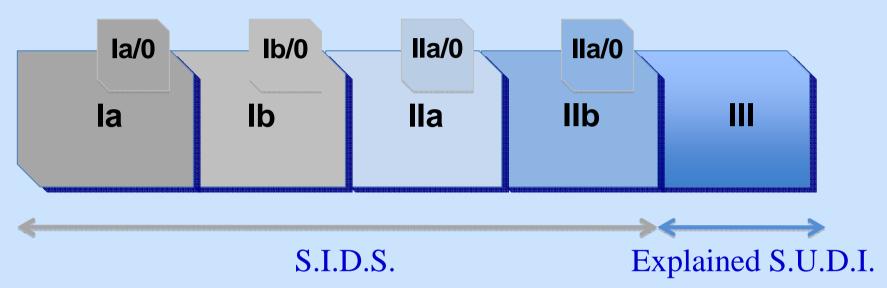
III: Factor(s) identified that **provide** a cause of death.

The 2007 Soria Moria course on SIDS and pediatric forensic medicine and pathology. Oslo, November 29-December 1, 2007

An expert panel that met in San Diego (2004) recognised the difficulty with incomplete investigations and how this may impact on a classification system.

How to distinguish between an incomplete investigation where a minor or major piece of information is missing?

They added a classification "0" which recognises such deaths as a S.U.D.I. but also recognises that a major pieces of information was lost and no decision could be made (explained or unexplained).



"0": Incomplete investigation (classified S.U.D.I.)

"'0": Extension used to denote that a potentially important piece of information is missing

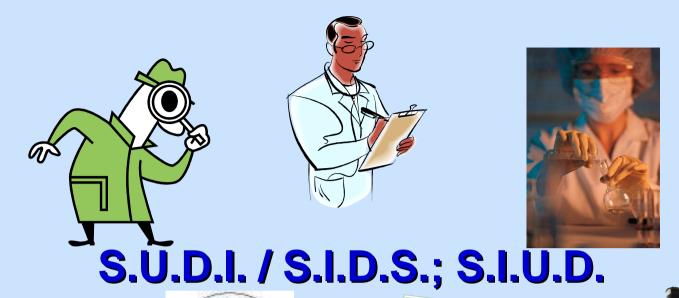
Ia: No notable factors identified

Ib: Notable factors identified but not likely to have contributed to the death

IIa: Factor(s) identified that possibly contributed to the death

IIb: Factor(s) identified that probably contributed to the death

III: Factor(s) identified that **provide** a cause of death.









a multi-agency approach









S.U.D.I. / S.I.D.S.; S.I.U.D. a multi-agency approach











Commissione Regione Toscana
per l'elaborazione dei percorsi organizzativi finalizzati
all'esecuzione del
riscontro autoptico nei casi di SIDS

(decr. 2011 del 12/05/2008)

