Sudden Infant Death Syndrome (SIDS)

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**SIDS** - **sudden death** of infant < 1 year that remains unexplained after thorough case investigation:

1. review of **clinical history** + infant and family medical and social history.
2. **examination of death scene** (esp. look for CO exposure, high ambient temperature, obstruction of external airways, accidental head entrapment).
3. **complete autopsy** (normal hydration and nutrition; no signs of obvious or occult trauma; *intrathoracic petechiae* are typically present on surfaces of thymus, pleura, and epicardium);
	* obtain whole-body ***radiographs*** (for evidence of skeletal trauma).
	* in many jurisdictions, ***toxicologic screening*** of serum and ***vitreous electrolyte analysis*** are routinely performed.

N.B. cases that fail to meet this definition (e.g. no postmortem investigation), should not be classified as SIDS! then diagnosis must be *sudden unexpected death in infancy* *(SUDI)*.

Typical history

* infants are typically born **full term** without history of significant pregnancy-related complications.
* no outward signs of significant health-related concerns are observed (**feeding well**, **gaining weight** **normally**).
* 70% infants have history of minor (!) viral upper respiratory tract or GI illness in week preceding death.
* infant is recently fed and then placed for sleep; when next checked, infant is discovered without pulse or respiration (with *frothy blood-tinged discharge* from nose or mouth, livor mortis and rigor mortis).
* 5% cases have history of **ALTE**\* preceding death.

\*old names - near-miss SIDS, aborted crib death

**ALTE (apparent life-threatening event)** - episode that is frightening to observer and is characterized by some combination of apnea (central or obstructive), change in muscle tone (usually diminished), and choking or gagging;

* frequency among healthy term infants is 1-3%.
* risk of subsequent death among infants experiencing ALTE is 1-2%.
* identifiable causes of ALTE: apnea of infancy, gastroesophageal reflux disease, respiratory syncytial virus bronchiolitis, pertussis, sepsis, meningitis, seizure, breath-holding spell, long QT syndrome, anemia, structural CNS anomaly, cardiac or airway anomaly.
* 50% of ALTEs remain unexplained!!!
* ***in-hospital observation*** is suggested for most infants following ALTE.

Epidemiology

SIDS is most common cause of death in postneonatal period (1 mo ÷ 1 yr) - causes 35-55% deaths.

* male-to-female ratio = 3:2
* 2/3 deaths occur in infants **2-4 months old** (90% < 6 months; 95% < 8 months); almost no cases are seen prior age 1 month.
* incidence (2002) - 0.51 per 1000 live births (represents 58% decrease from 1992 when "Back to Sleep" campaign was started and prone sleeping rate fell from 75% to 11.3%).
* *alternative diagnoses* are identified in 15-25% sudden unexpected deaths in infancy (SUDIs).

Risk factors:

* 1. sleeping prone!!!
	2. cigarette smoke exposure (principally in utero exposure) → 2- to 5-fold elevated risk.
	3. bed-sharing - infant may be suffocated by soft, loose bedding or sleeping adult.
	4. infant overbundling / overheating
	5. African American and Native American race → > 2-fold elevated risk.
	6. sibling with SIDS (risk increased 20-fold)

Factors that decrease risk:

1. room sharing (reduces risk by 50%)
2. pacifier (dummy) use - modifies cardiac autonomic balance during sleep.
3. Hispanic white race.

Etiopathophysiology

- triple risk model: Dysfunction of neural cardiorespiratory control!

1. **Vulnerable infant** (intrinsic abnormalities in cardiorespiratory control)
2. **Critical period** of development of homeostatic control mechanisms (persistent fetal reflex responses → enhanced inhibitory and depressed excitatory cardiorespiratory reflex responses to local stressors)
3. **Exogenous stressors** (e.g. changes in oxygen or carbon dioxide concentrations within sleep microenvironment, changes in ambient temperature, interactions with infectious agents).

Death occurs when vulnerable infants are subjected to stressors at times when normal defense mechanisms may be structurally, functionally, and/or developmentally deficient.

Numerous structural / functional neuro abnormalities have been described in infants with SIDS:

1. delayed development of brain stem (elevated dendritic spine counts, ↓degree of myelination)
2. differences in CNS dopamine beta-hydroxylase and tyrosine hydroxylase.
3. abnormalities in adrenergic pathways related to cardiorespiratory control within medulla and pons.
4. abnormalities of arcuate nucleus (nucleus is critical to integration of cardiorespiratory and arousal responses) - fewer muscarinic receptors, structural deficiency, reduced kainate binding.
5. defects in serotonin-binding neural pathways.
6. defective pathways within medulla, cerebellum, limbic system and prefrontal cortex.

Prevention

American Academy of Pediatrics "Back to Sleep" Recommendations:

1. place infant exclusively on back (supine) for every sleep; side sleep position is not as safe as supine and is not recommended.
2. infant must sleep in separate bassinet or safety-approved crib; use firm safety-approved crib mattress with tight fitting sheet; excessively soft or padded sleep surfaces (pillows, quilts, comforters, sheepskins) should not be placed under infant.
	* keep soft objects and loose bedding out of crib.
	* if blankets are used, they should be tucked in under mattress so that infant's head is less likely to become covered by bedding.
	* place infant so that its’ feet are positioned at foot of crib.
	* use sleepsack as alternative to blanket; make sure that infant’s head remains uncovered during sleep.
3. do not smoke during pregnancy + avoid second-hand smoke exposure for infant.
4. room-sharing sleep arrangement is recommended; bed-sharing is associated with higher risk.

***Cosleeping on couch or sofa*** is associated with unusually high risk for SIDS!

1. offer pacifier when placing infant down for sleep.
	* reinserting pacifier (if it falls out) is not necessary once infant is asleep.
	* do not force use of pacifier if infant refuses it; do not sweeten pacifier to enhance its use.
	* ***delay use of pacifier until after one month of age for breast-fed infants*** (to ensure establishment of breast-feeding).
2. avoid overheating and overbundling.
3. avoid commercial devices marketed to reduce SIDS risk; do not use home monitors as strategy to reduce SIDS risk.
4. avoid development of *positional plagiocephaly* - use "tummy-time" when awake.

Management

* autopsy should be performed quickly; preliminary results should be communicated to parents.
* parents are grief-stricken and unprepared for tragedy.
* because no definitive cause can be found, parents have excessive guilt feelings.
* family requires ***support for at least several months***.

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