



Sudden Cardiac Death In the Young - The First Few Hours

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Akron Children's Hospital

Why Am I Here?

- Sudden Death in the Young
- Duration of CPR, Outcomes, and the future
- Death Scene Investigation



911, How can I help you?

- All cause sudden death in children is rare (Wren, et al 2002, 2014)
 - 1:20k-50k children
 - 10% of all pediatric deaths
 - 50%
 - Epilepsy, asthma, cardiovascular abnormalities
 - 33% unexplained
 - Infection
 - Occult cardiovascular anomaly



Sudden Cardiac Death by mechanism

Situation/Lesion	Mortality Rate	Arrhythmia?	Comment
Perioperative	0.9/ 1000 patient-years	Yes, during terminal event and less common during life	Initially due to pulmonary artery hypertension
Tetralogy of Fallot	1.4/1000 patient-years	Yes, related to pulmonic regurgitation and RV dysfunction	Overall rare; young adult > children
Atrial repair of TGA	5-6/1000 patient-years	Sinus node failure after 10-15y	junctional rhythm common but seems unrelated to death
Adult CHDs	5.3/1000 patient-years	Yes	Eisenmenger's > TOF > TGA
Other congenital heart lesions	Rare event 0.1/1000 patient-years		
Hypertrophic cardiomyopathy	Unknown denominator		



Main Causes of Sudden Cardiac Death in Children

Primary arrhythmias

- ▶ Long QT syndrome
- ▶ Wolff-Parkinson-White syndrome
- ▶ Atrioventricular block
- ▶ Other ventricular arrhythmia (for example, catecholaminergic ventricular tachycardia, Brugada syndrome, etc)
- ▶ Non-hypertrophic varieties of hypertrophic cardiomyopathy (troponin T mutation)

Other structural heart abnormalities

- ▶ Hypertrophic cardiomyopathy
- ▶ Myocarditis
- ▶ Dilated cardiomyopathy
- ▶ Kawasaki disease
- ▶ Arrhythmogenic right ventricular cardiomyopathy

Main causes of sudden cardiac death in children

Cardiovascular malformations

- ▶ Atrial repair of transposition of the great arteries (Mustard and Senning operations)
- ▶ Repaired tetralogy of Fallot
- ▶ Aortic valve stenosis
- ▶ Coarctation of the aorta
- ▶ Pulmonary atresia
- ▶ Pulmonary vascular disease (Eisenmenger syndrome)
- ▶ Coronary artery malformations



Scene run

- Akron EMS called to home with 4 yo boy who was chasing his sister, stopped and fell to ground
- On arrival he is pulseless in a non-shockable rhythm. PALS is initiated and continues to the ED
- There is no ROSC after 45 minutes and all branches in the PALS algorithms have been explored and exhausted?
- How long should prehospital CPR be continued in children?



What do we know?

- Resuscitation duration
 - Professional CPR → ROSC
- ROC-PRIMED
 - Single blind, multi-centered, cluster randomization trial
 - Specific Aim: resuscitation duration and outcome
- Outcomes
 - Survival to hospital discharge
 - Favorable
 - Unfavorable
 - ROSC
 - Without survival
 - Never



Appropriate Duration of Resuscitation After Cardiac Arrest

Nov 17, 2016

Authors:

Reynolds JC, Grunau BE, Rittenberger JC, Sawyer KN, Kurz MC, Callaway CW.



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Appropriate CPR Duration after OHCA in Adults

(Reynolds JC et al 2016)

- 11,000 adults
 - Mostly males, 69yo (median)
 - Professional CPR duration 20 minutes (median)

• OUTCOME	Percentage	Professional CPR Duration (median)
• No ROSC	65% (7000)	23.4 minutes
• ROSC	35% (4000)	13.5 minutes
• Survive to DC	10% (1200)	
• Favorable	8% (905)	

- Favorable outcome declined with the duration of resuscitation
 - 37 minutes appeared to be a significant breakpoint and affected outcome
 - Favorable factors → ROSC + favorable outcome with <40 minutes of professional CPR
 - Initial shockable rhythm
 - Witnessed arrest
 - Bystander CPR



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What is the optimal duration of professional CPR after OHCA in adults?

The majority of patients with good neurological recovery after OHCA need CPR for ≤ 20 minutes, although there were a small number of patients who survived after prolonged cardiac arrest. An arbitrary cutoff for CPR duration (especially if short) might result in death of some patients who would otherwise survive with good neurological recovery.



Outcomes after non-traumatic OHCA in France (2017)

- Interaction between “down time” (No-Flow and professional CPR time (“Low Flow”))
- National resuscitation registry
 - 4.5 years to December 2015
 - Mostly adult men over 65
- Two tiered system (66M citizens)
 - Emergency Physician led team and/or Fire/EMT Team
 - All OHCA get BOTH
- Outcomes
 - 40,000 OHCA
 - 27,000 eligible
 - 14 minutes to professional CPR
 - NO-FLOW 10 minutes
 - LOW-FLOW 30 minutes
 - 26% ROSC (7300)
 - 20% survive to admission
 - 5% 30d survive to discharge (1480)
 - 4.5% favorable outcome (1250)

Cardiopulmonary resuscitation duration and survival in out-of-hospital cardiac arrest patients[☆]

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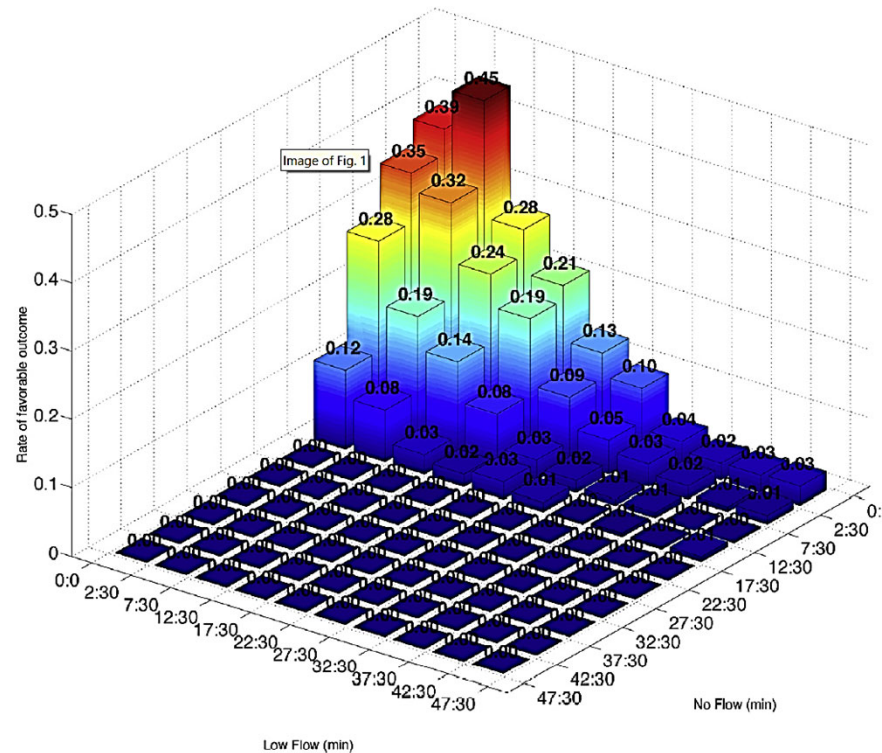
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No-Flow and Low-Flow Interaction and Outcomes-Overall



Conclusions

- There is significant interdependence between the No-Flow and Low-Flow interval and functional outcome after OHCA
- The cardiac arrest duration (NF+LF) associated with favorable outcome is adversely impacted as the NF interval increases
- Long duration of CPR is most effective (favorable outcomes) in cases of a shockable rhythm
- CPR in excess of 10 minutes in the cases of a non-shockable initial rhythm, even when witnessed is associated with very poor outcomes
 - Probability of survival without sequelae falls to < 1%
 - After 48 minutes with shockable initial rhythm
 - After 15 minutes with non-shockable initial rhythm



What about in children?

- Japan (20M children <17y)
- Utstein-style public registry
- Ambulance Crew
 - 3 EMT/Paramedics
- *“EMS providers are not permitted to terminate resuscitation in the field, excluding victims of decapitation, incineration, decomposition, rigor mortis, or dependent cyanosis. Thus, most OHCA patients treated by EMS personnel are transported to a hospital and included in the registry.”*

Shida et al. *Scandinavian Journal of Trauma, Resuscitation and Emergency Medicine*
(2019) 27:79
<https://doi.org/10.1186/s13049-019-0658-7>


Scandinavian Journal of Trauma,
Resuscitation and Emergency Medicine

ORIGINAL RESEARCH

Open Access

Prehospital cardiopulmonary resuscitation duration and neurological outcome after out-of-hospital cardiac arrest among children by location of arrest: a Nationwide cohort study



Haruka Shida¹, Tasuku Matsuyama^{2*} , Kosuke Kiyohara³, Tetsuhisa Kitamura⁴, Takefumi Kishimori¹, Takeyuki Kiguchi⁵, Chika Nishiyama⁶, Daisuke Kobayashi⁵, Satoe Okabayashi⁵, Tomonari Shimamoto⁵, Takashi Kawamura⁵ and Taku Iwami⁵



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Younger pediatric patients tend to have unwitnessed collapse at home

- 36m → December 2015
- 373k entered
 - 3900 children
 - 2865 residential
 - 1048 public
- Younger children
 - Residential areas
 - Medical causes
 - Bystander CPR
 - Unwitnessed
 - First rhythm non-shockable
 - Not public-access AEDs

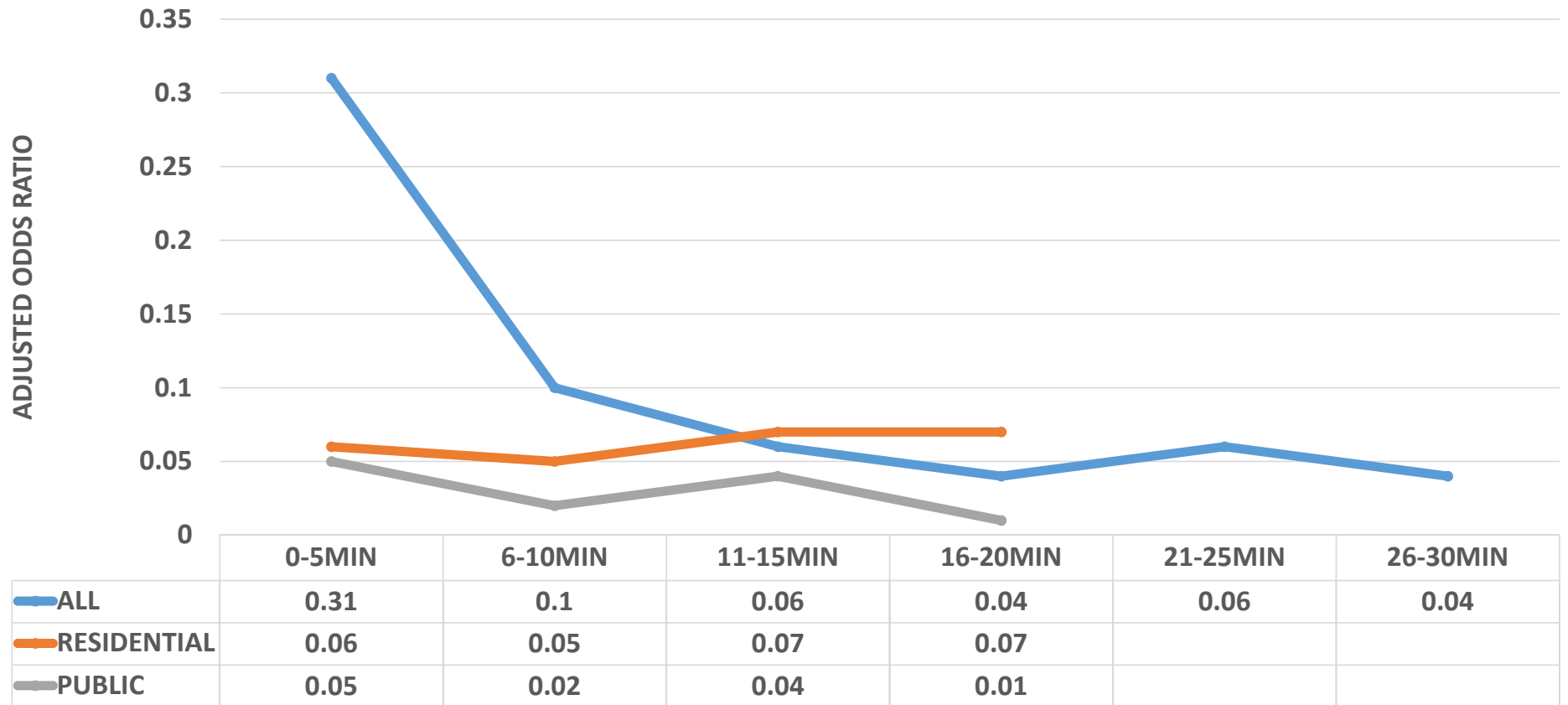


Primary Outcome: Favorable 30d neurologic outcome

LOCATION	PCT FAVORABLE	N
RESIDENTIAL, 2865	2.3%	66
PUBLIC, 1048	10.8%	113



CPR DURATION AND FAVORABLE 30D NEUROLOGIC OUTCOME P-VALUE WAS SIGNIFICANT FOR ALL TRENDS IN ALL GROUPS OVER TIME



Conclusions

JAPANESE UTSTEIN REGISTRY 2019

- Longer duration of prehospital professional CPR is independently associated with a increased proportion of patients having unfavorable 30d neurologic outcome
 - This effect is negatively modulated by collapse location
 - N.B. favorable 30d neurologic outcomes were reported after prolonged (>30min) professional CPR in both residential (1.4% [6/417]) and public collapse (0.6% [1/170])
- There is insufficient evidence to recommend a cutoff for individual patients
 - Many doubt such a recommendation is possible



Can we learn anything from Death Scene Investigations?

PEDIATRICS' PERSPECTIVES

A New Approach to the Investigation of Sudden Unexpected Death

Richard D. Goldstein, MD,^{a,b} Henry M. Nields, MD, PhD,^c Hannah C. Kinney, MD^d



METHODS AND PROCEDURES



**Improving Forensic Pathologic Investigation of Sudden Death in the
Young: Tools, Guidance, and Methods of Cardiovascular Dissection
from the Sudden Death in the Young Case Registry**

1. Goldstein RD, et al Pediatrics 2017;140(2):e20170024
2. Gulino SP, et al Acad Forensic Pathol 2018 8(2): 347-391



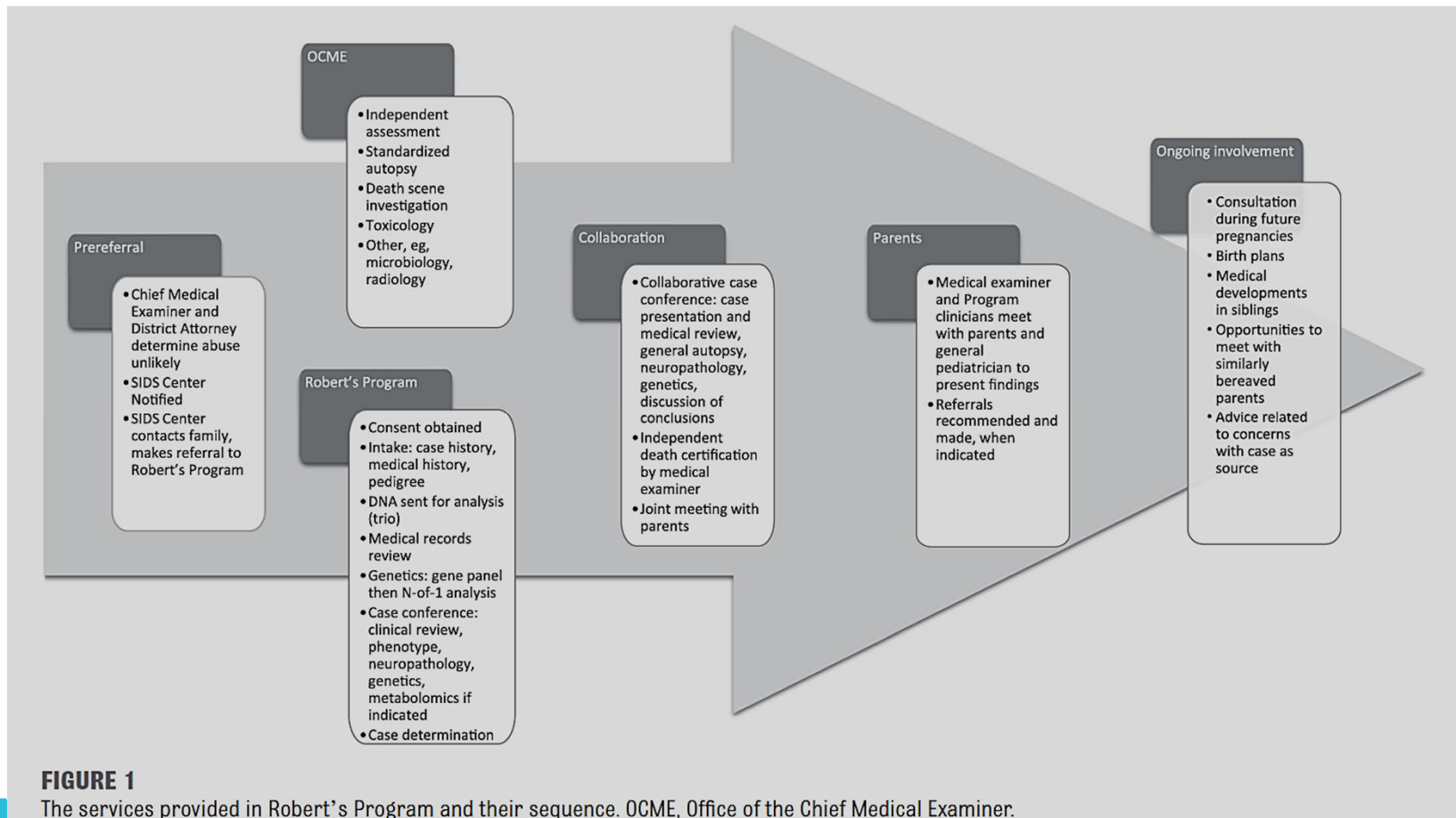
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Sudden Unexpected Death and Public Health Quality Improvement

- Sudden death is more likely in children < 19yo than cancer or cardiac death (CDC 1999-2014)
- Evaluation of the living is decreasing the percent of idiopathic cases
- Robert's Program (Boston Children's & Massachusetts OCME)
 - Multidisciplinary
 - SUDP is due to undiagnosed or undiscovered diseases in children < 3 yo
- Parents
- Boston Children's multidisciplinary clinical scholars
- OCME
- General Pediatricians



Robert's Program, a Team of Teams



Roberts Program-Collaboration

Robert's Program

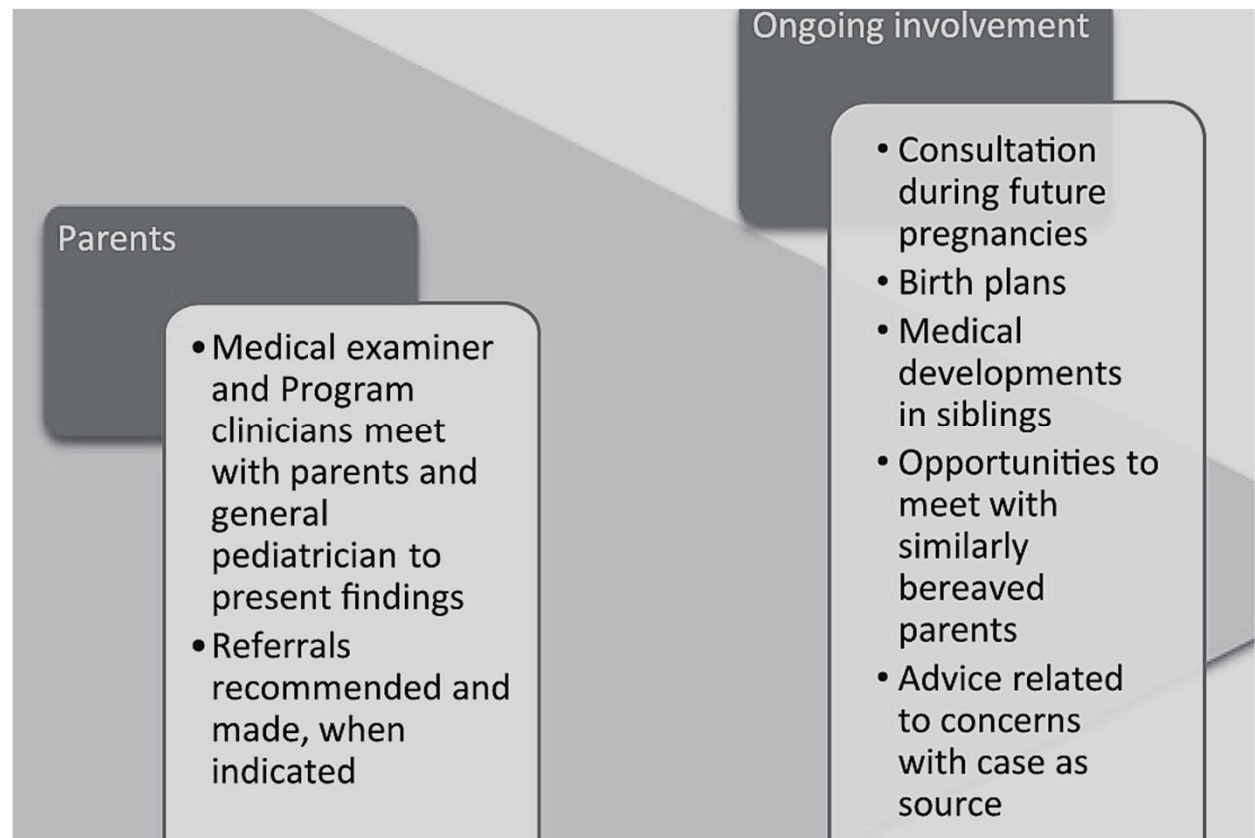
- Consent obtained
- Intake: case history, medical history, pedigree
- DNA sent for analysis (trio)
- Medical records review
- Genetics: gene panel then N-of-1 analysis
- Case conference: clinical review, phenotype, neuropathology, genetics, metabolomics if indicated
- Case determination

Collaboration

- Collaborative case conference: case presentation and medical review, general autopsy, neuropathology, genetics, discussion of conclusions
- Independent death certification by medical examiner
- Joint meeting with parents



Roberts Program- Next Steps



Brighter Future

- Family interviews
- Greater community involvement
- Greater collaboration



1. Gulino SP, et al Acad Forensic Pathol 2018 8(2): 347-391



METHODS AND PROCEDURES



Improving Forensic Pathologic Investigation of Sudden Death in the Young: Tools, Guidance, and Methods of Cardiovascular Dissection from the Sudden Death in the Young Case Registry



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Conclusions

- Sudden Death in the Young
 - More common than cardiac or cancer death
 - Less idiopathic deaths than ever
- Duration of CPR, Outcomes, and the future
 - Still unknown, but public collapse appears more favorable
- Death Scene Investigation
 - Team of Teams learning from the victims and survivors

