The following information is submitted to be included in the review of quality of care provided to [redacted] by Raymond Fripp (Control #800 2014 010003), Kathleen Kaya (#800 2014 010006), and John William Moore (#800 2014 010009)

The following list highlights a few complaints regarding Rowan's care. This list is not inclusive of all concerns, and should not be regarded as such:

1. **Current research states that anesthesia should be used only with great caution for patients with Williams Syndrome.** (see appendix A - References on sudden death as a result of Anesthesia in Williams Syndrome)

2. **Clear postoperative planning and early intervention are the hallmarks of life-saving care for patients with WS. The following table shows disparities that were found between recommended Williams Syndrome Protocol and the practices at Rady Children's Hospital:**

Williams Syndrome Recommended Protocol	Rowan's care at Rady Children's Hospital
Pre-Anesthetic EKG - *Ischemia from CA pathology is probably the most important reason for death*. It is imperative to obtain an EKG looking for signs ventriculomegaly, LV strain, ischemia, and infarction	Last EKG performed two months prior (10/21/13)
Pre-Anesthetic Echocardiogram - Echo helps define the location of SVAS, arch integrity, extent of pulmonary tree involvement, overall ventricular function, and areas of focal wall motion abnormality. *It is important to remember that gradients do not always correlate with the severity of CA pathology patients with relatively mild SVAS were later found to have severe CA abnormalities on autopsy.*	Last ECHO performed over one year prior (11/2012) (Fripp required that Rowan be sedated for the 11/2012 echocardiogram)
Pre-Anesthetic ICU admission for close hemodynamic monitoring is clearly indicated for patients who have baseline evidence of cardiac dysfunction. ECMO has been required in some cases to rescue children from total CV collapse. ***Thus, elective cases should be scheduled during times of maximum staff support, and arrangements for surgical/perfusion stand-by should be coordinated well in advance.***	Admitted as outpatient. ECMO brought in 25+ minutes post-anesthetic induction. Mother's request for inpatient status refused.
Consent - There is always the small possibility of respiratory and/or cardiovascular compromise during sedation or general anesthesia, ***but death is a very real component of care for patients with cardiac manifestations of WS. The risk of sudden death cannot be simply made on the basis of an ECHO study or otherwise.*** Although some patients demonstrate very obvious features of severe cardiovascular dysfunction and carry great	Parents were never informed of the possibility of sudden death

anesthetic/surgical risk, ***parents must understand in no uncertain terms that complete hemodynamic collapse and death can occur even in patients with "mild" disease.***	
NPO Status - Patients must maintain a consistent intravascular volume to preserve adequate LVEDV and subsequent LVED. It is important to encourage liberal fluid intake until shortly before the procedure per standard NPO guidelines. ***Early IV placement and correction of any outstanding fluid deficits is paramount.***	Parents were directed to withhold fluids, extra fluid intake not discussed with parents, IV placed after anesthetic induction
Induction - the goals for induction in a patient with WS are similar to those with severe AS: Maintain normal heart rate and rhythm, Maintain a high preload/optimize intravascular volume status, Avoid fluctuations in SVR and PVR, Avoid agents that cause myocardial depression	PVR and SVR not monitored, heart rhythm not monitored, used agents known to cause myocardial depression
Close post-operative monitoring is essential. Be it in the PICU or the PACU, the team of nurses and physicians must anticipate and correct any early signs of hemodynamic dysfunction. ***Resuscitation can be extremely difficult, if not impossible, once the patient arrests with severe SVAS and CA pathology.***	Unknown

(Source for Table: **A cautious approach to anesthesia for patients with Williams Syndrome.** *Anthony Clapcich. MD and Lynne Maxwell MD. Children's Hospital of Philadelphia.* *http://www.pedsanesthesia.org/meetings/2004winter/pdfs/PBLD3.pdf*)

3. **Rowan's mother and father voiced their concern about the use of anesthesia many times, over the telephone and in person**. The following samples indicate that this concern is referred to in Rowan's medical records:
   A. Dr. Lynne Bird, Rady Children's Hospital, November 2012 wrote: (In regard to an MRI) Rowan's parents "are reticent about the anesthetic risk..."
   B. Dr. Raymond Fripp, May 2013 wrote: "… (A CT scan) will require general anesthesia and Rowan's mother has some reluctance to him having GA..."
   C. Rowan's mother, December 2013 wrote on the "Acknowledgement of Risks of Anesthesia" form: "*Signed with the expectation that the anesthesiologist is well-versed in the increased anesthetic risk associated with patients who have William's Syndrome TMG 12/16/13 6:45 am*"

4. **Rowan's mother and father were never told of any risk to Rowan's life as a result of anesthesia**
   A. This is documented by its omission in the medical notes prior to Rowan's death

5. **Rowan's mother and father were not given any other options for diagnostics outside of an anesthetized CT angiogram**. This omission is noted in Rowan's medical records here:
   A. Dr. Raymond Fripp, October 2013 wrote: "I once again had a prolonged discussion with Rowan's parents regarding the need for a CT angiogram to assess the coronary arteries and make sure there are no coronary artery stenosis which is not uncommon in children with Williams syndrome. The CT will also allow us to look at the anatomy of the branch pulmonary arteries and the supra valve aortic area."

   i. Subsequent to Rowan's death, we located information about the following alternatives (none of which was offered to, or discussed as, an option for Rowan):
      a) A functional stress-test, used in place of a CT angiogram for heart imaging
      b) Play therapy, used in allowing for (MRI/CT) diagnostics of 60 to 90 minutes without sedation. The following examples show an un-sedated MRI success rate of up to 96 percent when using play therapy: "Reviewing the process of preparing children for MRI" Pediatric Radiol (2008) 38:271 and "Paediatric MRI under sedation: Is it necessary?" Pediatr Radiol (2011) 41:1353)
      c) An un-sedated echocardiogram
      d) Additional diagnostics, used prior to use of anesthesia for patients with Williams Syndrome (see above table)
   ii. Provision of, or parents' knowledge of, any of the above options would have allowed for safe cardiac diagnostic imaging as early as May 2013

When Rowan's family received a copy of his medical records, it was noted that the following information is incorrect:

1. **Incorrect information recorded after death:**
   After Rowan's death, additional medical records noted that Rowan was undergoing a diagnostic exam for SVAS. However, Rowan's parents and Dr. Raymond Fripp discussed several times that the CT angiogram was being performed to assess coronary artery stenosis. This was referred to in the medical records numerous times, including:
   A. Dr. Lynne Bird – July 25, 2013 wrote: Rowan "was seen again by Dr. Fripp… who would like to get a cardiac CT to evaluate the coronary arteries"
   B. Dr. Raymond Fripp, October 21, 2013 wrote: "I once again had a prolonged discussion with Rowan's parents regarding the need for a CT angiogram to assess the coronary arteries and make sure there are no coronary artery stenosis which is not uncommon in children with Williams syndrome."

   Coronary artery stenosis is a known significant risk factor for sudden death under anesthesia, while SVAS is not

2. **Incorrect information on medical record. On two occasions, Dr. Raymond Fripp referred to Rowan as "uncooperative" without justification:**
   A. Dr. Raymond Fripp, 10/21/13 – "From PHYSICAL EXAMINATION: …blood pressure 127/55 in the right arm with the patient uncooperative…" Dr. Fripp was not present when blood pressure was assessed. Rowan's mother witnessed Rowan cooperatively allowing the nurse to obtain BP reading, then leave to speak to Dr. Fripp, then return to take BP again. (see figure 1)
   B. Dr. Raymond Fripp, 12/12/13 note to pediatrician (Dr. Raul Sepulveda) - "Rowan is cleared from a cardiac standpoint to undergo general anesthesia for the CT. He will also have a sedated echocardiogram at the end of the CT since he is not cooperative enough to have an echocardiogram without sedation." (see figure 2)

   Rowan had visits with four different doctors and five different therapists. No similar notes were found in their records. On the contrary, the following is a sample from their notes:

   i. 5/8/13: Laura Boyer PT , Rady Children's Hospital wrote: "ASSESSMENT: … Rowan very smiley and cooperative."
   ii. 5/21/13: Jennifer Boche, DPT wrote at initial meeting "ASSESSMENT: Rowan is a friendly 20-month old little boy… He engaged readily with the evaluator."
   iii. 7/13 – Rowan underwent a diagnostic abdominal ultrasound with no sedation or anesthesia at Rady Children's Hospital
   iv. 8/3/13: Jenny Boche, PT wrote – "Rowan is very friendly and engaging. He responds well to peer-modeling and play-based intervention….. Rowan is able to recover quickly from frustrations"
   v. 10/21/13: Rowan sat cooperatively for an ECG (see figure A)
   vi. 10/30/13: Ayala Weinstain, OT wrote – "..he was very happy and engaged well.."
   vii. 11/22/13: Tiffany Harrison, Speech wrote – "..he participated well in session.."

3. **Missing information and discrepancy of documentation between a) the time of admission and time of anesthetic induction, and b) the initiation of CPR, involvement of advanced medical team**
   A. The time (prior to initiation of CPR) documented on Rowan's medical records does not coincide with the time documented by parents (documented on telephone and email records, available on request)
   B. A significant time lapse is noted between the time of cardiac arrest and the involvement of the advanced medical team and William's Syndrome appropriate medical interventions

The complaint to the Medical Board is concerning only the time up until the initiation of CPR. **We are not concerned with the medical interventions that took place subsequent to that event.**

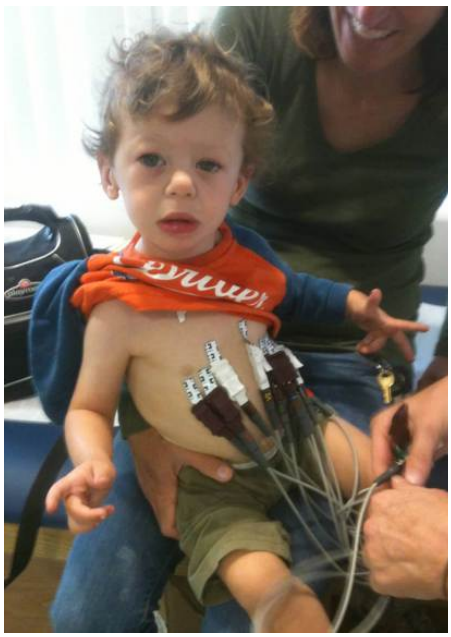

Figure 1 - Rowan receiving an EKG at Rady on 10/21/2013

Figure 2 - Rowan and his sister at Rady Children's Hosp 12/12/13

Appendix A - References on sudden death as a result of Anesthesia in Williams Syndrome:

1. **Gupta et al., 2010. Annals of Cardiac Anesthesia: Sudden cardiac death under anesthesia in pediatric patient with Williams syndrome: a case report and review of literature.**

Abstract: Williams syndrome is a complex syndrome characterized by developmental abnormalities, craniofacial dysmorphic features, and cardiac anomalies. Sudden death has been described as a very common complication associated with anesthesia, surgery, and procedures in this population. Anatomical abnormalities associated with the heart pre-dispose these individuals to sudden death. In addition to a sudden and rapid downhill course, lack of response to resuscitation is another significant feature seen in these patients. The authors report a five-year-old male with Williams syndrome, hypothyroidism, and attention deficit hyperactivity disorder. He suffered an anaphylactic reaction during CT imaging with contrast. Resuscitation was unsuccessful. Previous reports regarding the anesthetic management of patients with Williams are reviewed and the potential for sudden death or peri-procedure related cardiac arrest discussed in this report. The authors also review reasons for refractoriness to defined resuscitation guidelines in this patient population.

Summary: The authors conclude from a detailed case report and literature review that sudden death as a result of cardiac arrest under anesthesia is a significant risk factor for individuals with WS. The also conclude that pre-operative identification of at-risk WS patients is very difficult. **Consequently, very detailed risk-benefit analysis should be done before any elective diagnostic procedures requiring anesthesia, and parents should be fully informed of the potential risk of sudden death as a consequence of anesthesia.**

2. **Olsen et al., 2014. Anaesth. Intensive Care. Anaesthesia-related haemodynamic complications in Williams syndrome patients: a review of one institution's experience.**

Abstract: Williams syndrome is a genetic disorder associated with cardiac pathology, including supravalvular aortic stenosis and coronary artery stenosis. Sudden cardiac death has been reported in the perioperative period and attributed to cardiovascular pathology. In this retrospective audit, case note and anaesthetic records were reviewed for all confirmed Williams syndrome patients who had received an anaesthetic in our institution between July 1974 and November 2009. There were a total of 108 anaesthetics administered in 29 patients. Twelve of the anaesthetics (11.1%) were associated with cardiac complications including cardiac arrest in two cases (1.85%). Of the two cardiac arrests, one patient died within the first 24 hours postanaesthetic and the other patient survived, giving an overall mortality of 0.9% (3.4%). We conclude that Williams syndrome confers a significant anaesthetic risk, which should be recognised and considered by clinicians planning procedures requiring general anaesthesia.

Summary: The authors perform a detailed retrospective case review covering 5 decades of WS patient history at their institute. They conclude that serious cardiovascular complications as a direct result of anesthesia, including sudden death, are significant causes of morbidity and mortality. They also conclude that identification of "at-risk" WS patients is very difficult, so all WS individuals must be treated as high risk for cardiovascular complications and sudden death from anesthesia. Children under the age of 5 were also more at risk that younger cohorts. Consequently, **all anesthesia and sedation should be carefully considered and avoided if at all possible due to the apparent minimal cardiac reserve, especially in younger children.** The benefits of any proposed procedure should very carefully be weighed against the significant risk and the parents counseled in detail about the risk of sudden death and serious cardiac complications.