Angelman syndrome is a complex genetic condition characterized by ataxia, seizure disorder, characteristic EEG, subtle dysmorphic facial features, severe learning disabilities and a happy, sociable disposition. Angelman syndrome is caused by a deletion of chromosome 15q11-13, and is an example of an imprinting disorder. The genes located within this deleted region of chromosome are known to include a cluster of genes encoding the gamma-aminobutyric acid receptor (GABA) subunits. Many actions of our general anesthetic agents are mediated through the GABA-A receptor, a ligand-gated chloride channel, and act by enhancing receptor sensitivity to GABA and by prolonging chloride conductance. Therefore it has been speculated that Angelman syndrome patients have an abnormal response to GABA-enhancing agents such as propofol, inhalation agents, and benzodiazepines. Some authors have suggested that children with Angelman Syndrome are sensitive to GABA binding agents and require less drug than those with normal receptors.

Our hospital (unnamed) is one of five hospitals enrolled in the Angelman Syndrome Natural History Study. Using the data collected from these patients during the perioperative period we have been able to perform a retrospective study and analyze if children with Angelman syndrome appear to be more sensitive to propofol or inhalation agents than other populations. The data studied included information from thirteen anesthetics performed in the last five years and contained children over a wide age-range from 12 weeks old to 9 years old. These patients represented many spectrums of the disease. Regardless of history of seizures, age, or the use of GABA activating drugs there were no complications seen in the perioperative period. Thus in the patients we reviewed undergoing general anesthesia at (institution name) we found no data to suggest that these patients demonstrate exaggerated responses to GABA stimulating drugs. In fact, despite proclaimed differing GABA receptor sensitivities, it appears that regardless of the anesthetic agent, there were no perioperative implications of Angelman syndrome.
