Advancements in the diagnosis and treatment of congenital heart disease has led to an increased population of patients who reach adolescence and adulthood who later require non-cardiac surgery. Many of these patients undergo correction that results in normal cardiovascular physiology, however a large subset of these patients undergo palliative surgery and exhibit abnormal cardiovascular physiology and function. This creates significant anesthetic challenges.

Unfortunately CHD itself, as well as the surgical treatments, have been associated with the development of scoliosis. While idiopathic scoliosis is estimated to occur in 2-3% of the general population, the prevalence is increased by both the presence of congenital heart disease as well as a history of sternotomy and/or thoracotomy. Scoliosis presents a significant source of morbidity for patients with CHD, as it can significantly impair respiratory function and therefore merits surgical correction. These surgeries are associated with increased blood loss in patients with normal cardiovascular physiology, and this is exaggerated in patients exhibiting Fontan physiology. The chronic increase in CVP can lead to abnormal coagulation due to hepatic congestion as well as increased venous bleeding during the procedure. While the use of anti-fibrinolytic therapies has been discussed to potentially decrease intra-operative blood loss, there are no published reports of its use in patients with CHD in the literature. We report a 17-year-old male with tricuspid atresia and hypoplastic right ventricle status post Fontan procedure who presented for T4-L2 posterior instrumentation and fusion to whom we administered peri-operative tranexamic acid.

Our primary concern was maintaining intravascular volume that provided adequate pulmonary blood flow and cardiac output. Secondarily, in an effort to decrease intraoperative blood loss, we chose to administer tranexamic acid during the case. Anti-fibrinolytic therapy has been suggested in the literature, however its use hasn’t been reported. Concerns about sluggish pulmonary blood flow and therefore an elevated risk for thromboembolism exist in patients with Fontan physiology. Our patient, however, had normal pre-operative coagulation studies and was only taking Aspirin 81mg as an outpatient. Both these factors made us more comfortable administering anti-fibrinolytic therapy. The eleven level fusion took six hours and 17 minutes and the patient experienced estimated blood loss of 1000 mL (27ml kg-1). He was extubated on post-operative day one and discharged from the hospital after an uneventful hospital stay on post-operative day three.

Rafique MB, Stuth EAE, Tasssone JC. Increased blood loss during posterior spinal fusion for idiopathic scoliosis in an adolescent with Fontan physiology. Paediatr Anaesth 2006; 16(2): 206-212