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Journal of Brain and Neurological Disorders

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Case Report

Thrombolysis with Intra-Arterial T-Pa for Acute Ischemic Stroke in Children Treated with Fontan Operation, Case Report

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Received Date: January 14, 2022 | **Accepted Date:** May 12, 2022 | **Published Date:** May 28, 2022

Citation: Mohammed Habib, Mohammed Hillis, Ala Alwihidy, Basel Oda. (2022). Thrombolysis with Intra-Arterial T-Pa for Acute Ischemic Stroke in Children Treated with Fontan Operation, Case Report. *Brain and Neurological Disorders*. 5(1); DOI:10.31579/2642-9730/015

Abstract

We report the case of 7-year-old women with acute ischemic stroke, previously treated with Fontan operation due to complex congenital heart disease at the age of 2 years, who underwent an intra-arterial thrombolysis with a favorable outcome. No controlled randomized trials on thrombolysis among childhood stroke have been conducted yet. This and other similar cases suggest that thrombolysis may also be safe and beneficial in pediatric patients.

Keywords: thrombolysis; acute ischemic stroke; intra-arterial t-pa

Introduction

Complex congenital heart disease such as single-ventricle in pediatric patients with requiring the surgical diversion of the systemic venous return from the superior vena cava (Glenn operation) and then the inferior vena cava (Fontan operation) directly to the pulmonary arteries. Because many of these patients are on chronic anticoagulation therapy and may have right-to-left shunts and arrhythmias disorders that predispose them to bleeding and/or clotting and casus acute cerebrovascular events (acute ischemic or hemorrhagic stroke). Thrombosis remains a major problem among pediatric patients with Fontan for a few reasons. Stasis and polycythemia from chronically low oxygen saturations is common and leads to increased blood viscosity predisposing to thromboembolic events [1]. One retrospective study showed an overall occurrence rate of 3.9 events per 100 patient-years, with an overall mortality rate of 21 % in those with a thrombus [2]. another trial suggested (4.2%) developed stroke over 5 years follow-up period and 75% of them within 2 weeks of surgery [3]. One study showed that Fontan patients on an antiplatelet or anticoagulant had lower rates of death compared to those who were not [4-5]. In this case we describe a 7-year-old woman born with complex congenital heart disease and operated with Fontan operation presents with acute ischemic stroke and treated successfully by mechanical thrombectomy and intraarterial t-PA.

Case report

A 7-year-old female (21 kg, 123 cm, BMI 16.9) while she was playing with her siblings presented to the emergency department with facial weakness, slurred speech, right arm and right leg weakness. Her last known normal 5 hours prior to arrival. No history of head trauma. Emergency Medical department were notified and the patient was transferred to do urgent brain CT. The patient was born with complex congenital heart disease; mitral valve atresia, double outlet right ventricle (DORV), Transposition of great arteries, large Ventricle septal defect, hypoplastic Lt ventricle like single ventricle and Pulmonary stenosis. The patient's medications included 81milligrams aspirin every other day. Vital signs on arrival were a blood pressure of 115/60, heart rate of 84 beats per minute, respiratory rate of 21 breaths per minute and oxygen saturation of 93% on room air. The patient history cardiac surgery BT Shunt at 4/2014, Glenn at 12/2014 and Fontan at 6/2016.

On exam the patient was resting comfortably and in no acute distress. she was complete aphasia left upper and lower facial droop. she was unable to move his entire left upper extremity, 0/5 in strength. Her left lower extremity was weak 2/5 in strength. The patient's NIH Stroke Scale was 11 for left sided facial palsy, left upper extremity paralysis, left lower extremity weakness, and aphasia.

Our patient underwent prompt neuro-imaging after establishing vascular access and documenting a blood glucose of 97. CAT scan of her head without contrast showed no intracranial hemorrhage no tumor and ASPECT score 8.

Selective left carotid angiogram confirmed complete occlusion of M1 segment of left MCA with an intraluminal thrombus (figure 1).

Figure 1: *MCA proximal totally occluded*

Figure 2: *opening of MCA after IA t-PA*

Thrombolytics such as tPA is recommended as long as the patient does not have any absolute contraindications. However, one of the primary indications
to administer tPA is age > 18 years old and within 4.5 hours from symptoms. If a child is suffering an ischemic stroke, tPA use would be off-label and treatment should be coordinated with pediatric sub-specialists. Management is based on clinical experience of experts, recommendations of consensus guidelines, case studies and extrapolation from adult clinical trial.

The patient underwent an IA alteplase 6 mg slowly over 15 minutes where the clot was successfully resolved from the left MCA with microcatheter. Post-thrombolytics angiography showed satisfactory flow in the left MCA. (figure 2).

The patient was admitted to the ICU where his neurological exam significantly improved during her hospital stay. He regained 5/5 strength in all extremities and his aphasia resolved but a mild left lower facial droop was still slightly visible when he smiled.

Discussion

Pediatric strokes are a relatively rare (1 to 2.5 per 100,000 children in the United States per year) compared to adult strokes (up to 970 per 100,000 adults between ages 65 and 74 in the Unities States per year) [6]. While strokes in adults are commonly linked to risk factors such as hypertension, hyperlipidemia, and diabetes, pediatric stroke risk factors include congenital heart disease, sickle cell disease and leukemia [7]. The patient in this case study had a history of complex congenital heart disease and operated with three-stage procedure that included the BT shunt, Glen, and Fontan surgeries.

Initial cohort studies reported the incidence of stroke from 3% to 19% in children treated with different thromboprophylaxis regimens. In 1,006 survivors of the Fontan operation, the Australia and New Zealand Fontan Registry reported an overall freedom from thromboembolism of 82% at 25 years after the Fontan procedure (95% CI,74–87%), with events occurring at a median of 7.6 years. In a subgroup analysis, freedom from thromboembolism in extracardiac Fontan patients was 91% at 12 years. Lateral tunnel Fontan patients had a thromboembolism incidence of 16%. The highest risk of thrombosis occurs in the first year after the Fontan operation [8].

Pediatric patients with Fontan operation are at particularly higher risk of systemic thromboembolic events such as stroke. The reported mortality from thromboembolism was 25% and death occurred despite aggressive therapy. The prevalence of thromboembolic complications in the literature is reported to be anywhere between 4% and 20%. Some studies have suggested dual peaks for thromboembolic events–the first within 12 months of operation and a second peak 10 years after the Fontan operation [3].

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DOI: 10.31579/2642-9730/015