

Short communication

Clinically successful late recanalization of basilar artery occlusion in childhood: What are the odds? Case report and review of the literature

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Abstract

Background and purpose: Acute basilar artery occlusion is an infrequent but potentially fatal cause of stroke, both in adults and children. We present our experience with a 6-year-old child and we investigate the rationality for late treatment of acute basilar occlusion in children.

Methods: We report the case of a 6-year-old boy with acute basilar artery occlusion presented with a full blown locked-in syndrome, admitted to the endovascular suite 44 h after the stroke onset, and we review all the reported cases of basilar artery occlusions presented with locked-in syndrome in children.

Results: Six hours following admission the basilar artery was partially recanalized by intra-arterial thrombolysis combined with mechanical clot angioplasty. After 12 h, the patient was awake, oriented, his speech function was fully restored and he had only a mild right hemiparesis that recovered completely after a month.

Conclusions: To our knowledge, this is the first report of complete clinical recovery after delayed (50 h) endovascular recanalization of basilar artery in a child. Intra-arterial thrombolysis combined with cerebral angioplasty, can successfully restore the patency of the basilar artery and the neurologic deficit of children with acute basilar artery occlusion, even after a considerable delay.

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1. Introduction

Acute basilar artery occlusion (ABAO) is an infrequent but potentially fatal cause of stroke, both in adults and children [1]. Locked-in syndrome is a common clinical pattern of this condition and is attributed to the occlusion of the midportion of the basilar artery (BA) causing ischemic damage to the pontine pyramidal tracts [2]. It is characterized by upper motor neuron quadriplegia, paralysis of lower

cranial nerves, bilateral paresis of horizontal gaze and anarthria, with preserved consciousness.

Traumatic vertebral artery dissection is one of the most common causes of ABAO in young patients and must be especially suspected in patients presenting with cervical pain preceding the dramatic neurological deterioration [3]. Delay in the diagnosis of ABAO is frequent due mainly to the misleading symptoms and signs and the rarity of this condition [4,5]. In adults, intra-arterial fibrinolysis is considered as the most effective available treatment to achieve BA recanalization and clinical improvement. In children, this technique has been rarely reported. The aim of this paper is to report the first case of complete clinical

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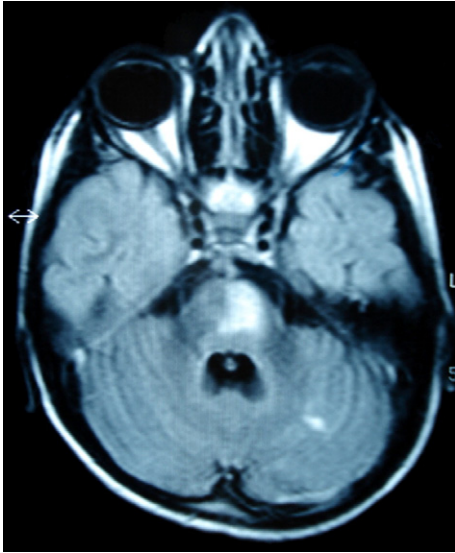


Fig. 1. Axial FLAIR shows acute left pontine infarction with lack of basilar artery flow void.

recovery after delayed (50 h) endovascular recanalization of the BA in a child and to review the pediatric literature.

2. Case report

A 6-year-old child presented an episode of sudden loss of consciousness a few minutes after the arrival in his classroom. This initial attack was characterized by an unprovoked shout followed by prolonged “pathological cry,” fall to the ground and “spasms”. The first assessment took place at the regional hospital and involved a neurological examination from a pediatric neurologist. On examination, the patient’s eyes were

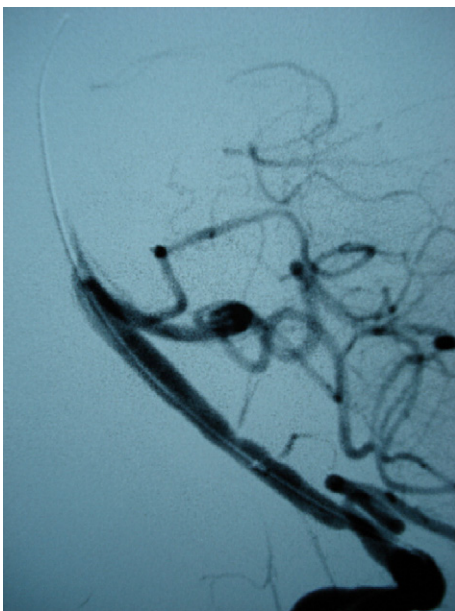


Fig. 2. Lateral left vertebral artery angiogram shows occlusion of the midbasilar artery, note the guidewire traversing the thrombus.

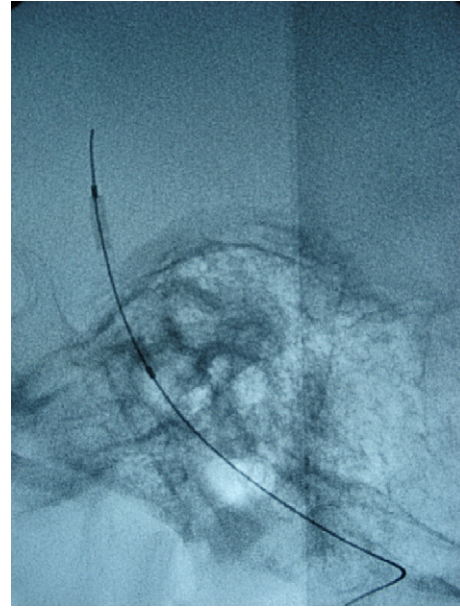


Fig. 3. Clot angioplasty.

open but there wasn’t any reaction to the environmental stimuli, he was aphasic and he had right hemiparesis. He underwent an urgent CT-scan which was informed normal. The patient’s medical history was unremarkable, except from a “minor” head injury 1 week before his admission while he played football. The admission diagnosis was “complicated epileptic seizure” and the decreased consciousness level of the patient was attributed to “post-ictal confusion”. Thus, he remained in the hospital for observation under anti-convulsive medication and continuous monitoring, and he was scheduled for an elective MRI investigation the next day. Twenty-six hours later, during his transfer to the MRI suite, the patient presented a second episode of generalized spasms. Neurological reevaluation revealed flaccid quadriplegia and mutism. He was hemodynamically stable and had open eyes with preserved blinking and vertical gaze. He was intubated and



Fig. 4. Anteroposterior postprocedural angiogram shows recanalization of the basilar artery and its branches.

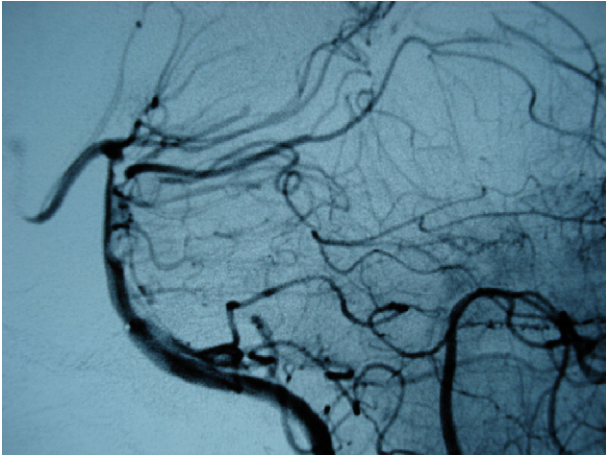


Fig. 5. The lateral postprocedural angiogram confirms the recanalization.

an urgent cerebral MRI/MRA performed. MRI disclosed a pontine infarction in relation to basilar artery occlusion (Fig. 1). The patient was admitted to the endovascular unit after 44 h of stroke onset.

2.1. Endovascular procedure

Under general anesthesia a 6F introducer sheath was placed in the right femoral artery. Selective 4-vessels cerebral angiogram was performed. Angiogram of the left vertebral artery revealed a complex dissection of the vertebral artery at the upper cervical level and complete BA occlusion (Fig. 2). The clot occluded the BA at the lower third and extended through the mid and upper basilar thirds. Heparin was administered intravenously (2000 UI) and activated clotting time was being controlled every 30 min. An Envoy (Cordis Endovascular) 6 F guiding catheter was placed at the proximal left vertebral artery. Coaxially, a microsystem composed by a microcatheter 2.7 F (Excel, Boston Scientific) and a 0.014-in. microguidewire (Transend, Boston Scientific) allowed selective catheterization of the BA. Intra-arterial infusion of 200,000 units of urokinase for 30 min allowed partial recanalization of the BA. Mechanical disruption of the clot was achieved by crossing the occluded artery with the microsystem. In order to improve the angiographic results, balloon clot angioplasty was performed with a semi-compliant angioplasty balloon 1.5 mm in diameter by 1.5 cm in length (Maveric, Boston Scientific) (Fig. 3). Partial BA recanalization was achieved but the main collateral branches and the brainstem perforating branches reappeared on angiography (Figs. 4 and 5). The patient was extubated 12 h after the end of the procedure and anticoagulation was initiated. ICU management included strict control of normoglycemia, pain therapy and avoidance of hypertension. He was hemodynamically stable, oriented, with normal speech and mild right hemiparesis (muscle strength 4/5). During his hospitalization (7 days) no complication occurred and his hematological and biochemical profile was normal. Anticoagulation was administered for 6 months, followed by

aspirin 75 mg per day. One month after the endovascular procedure his muscle strength in both right arm and leg was fully restored. He remained so during the next two follow ups, 3 and 9 months later.

3. Discussion

In general, the annual incidence of pediatric stroke is estimated at 2.5 cases per 100,000 children [6]. Furthermore, the occlusion of BA in childhood is even rarer [7]. Our review of the literature yielded 14 cases of ABAO presented with locked-in syndrome in children between 4–18 years of age (Table 1).

One of the leading causes of BA occlusion in children is the vertebral artery (VA) dissection [3]. Very often a history of trivial neck or head trauma is identified and this was the case in our patient. In adults, the mechanism of ABAO after a VA dissection is probably embolic, although others believe that it is the result of the intra-basilar extension of the initial dissection [8]. The dissection of VA in our case was located at V2 segment, which actually is the most often reported site.

The site of BA occlusion in the 14 children found in the literature with ABAO and locked-in syndrome was the mid-third of the BA in 6 cases [4,13–15,19], the proximal third in 2 cases [21,22] and the distal third in 1 case [5]. In 4 patients there was complete BA occlusion [12,14,18,23] and in 1 the location was not specified [20]. In our case, the occlusion site was the mid-third of the basilar trunk.

In adults, the site of BA occlusion has been correlated with the outcome: More distal occlusions have less favorable outcome [9]. When the occlusion is located at the proximal part of BA, there is a better chance for good outcome mainly due to the preservation of blood flow into the brainstem [9]. A possible mechanism for this, may be the collateral supply by the anterior circulation via the posterior communicating arteries. Taking into account that a similar collateral pattern was demonstrated in our case also, it is reasonable to hypothesize that this mechanism was, at least partially, involved in extending the therapeutic window.

In adults, BA occlusion is a devastating event carrying a fatality rate up to 83–92% if untreated [10]. Data collected from 12 different studies show that the functional outcome in adults is strongly related to the recanalization state, complete or partial. The rate of a good outcome without recanalization was approximately 2%, whereas in cases with some degree of BA patency this rate was risen up to 38% [11]. Preoperative factors related to the functional outcome include: initial clinical status, timing of intervention, location and length of occlusion, etiology, age and degree of collateral circulation [1]. Recanalization is the principal therapeutic goal in BA occlusion and intra-arterial fibrinolysis is the most effective therapeutic option. Only 5 of the 15 pediatric patients found in our review had received primary endovascular treatment [5,12–15].

By reviewing the clinical presentation of the patients that eventually developed locked-in syndrome, we concluded that

Table 1
Reported cases of BAO with locked-in syndrome in children: delay in diagnosis, treatment modalities and outcome

Study	Age	Site of occlusion	Delay from onset to treatment	Treatment	Outcome
1 Golden et al. [4]	13	Mid-BAO	Not specified	Not specified	Locked-in 18 months later
2 Larnar et al. [5]	18	Distal BAO	12 h	Intra-arterial thrombolysis; Heparin IV	18 months later: supervised walking, mild dysarthria, normal affect
3 Cognart et al. [12]	8	Complete occlusion	36 h	Balloon angioplasty; intra-arterial thrombolysis	Discharged with left hemiparesis and complete palsy of the vocal cords. At 3-month follow-up, normal
4 Kirton et al. [13]	15	Mid-BAO	20 h	Intra-arterial t-PA; balloon angioplasty	Remained locked-in for 24 h; improvement over the 1st week; at 6-month follow-up, mild incoordination of fine motor movements in the left hand; at 12 months, normal
5 Rosman et al. [14]	6	Mid-BAO	13 days	No anticoagulation; no antiplatelets; no thrombolysis	Locked-in for several months; restored speech; 4 years later, quadriparesis
6 Rosman et al. [14]	18	Mid-BAO	3 days	Intra-arterial t-PA	Locked-in 2 years later
7 Rosman et al. [14]	4	Nonfilling BA (MRA)	3 days	Heparin IV followed by warfarin	Locked-in for 3 weeks. 2 years later, left hemiparesis and ataxia
8 Zaidat et al. [15]	16	Mid-BAO	20 h	Clot retrieval; balloon angioplasty	Post-procedure: dysarthric but comprehensible; required quad cane for walking. At 3-month follow-up: minimal dysarthric speech, emotional lability, able to walk without assistance
9 Ackerman et al. [18]	10	Complete BAO	Not specified	Steroids	Mild quadriparesis 1 month later
10 Chatkupt et al. [19]	8	Mid-BAO	Not specified	Not specified	Locked-in 3 weeks later
11 Cheng et al. [20]	8	Not specified	Not specified	Anticoagulation; antiplatelets	Not positive (no more details given)
12 Latchaw et al. [21]	7	Proximal BAO	Not specified	Not specified	4 1/2 years later, locked-in with spastic quadriparesis
13 Mares et al. [22]	7	Proximal BAO	<24 h (not specified)	Heparin IV	Locked-in until death 4 weeks later from massive tracheal hemorrhage
14 Nakatomi et al. [23]	7	Complete BAO	24 h	Not specified	Paraparesis and CN XII palsy 10 months later
15 Grigoriadis et al.	6	Mid-BAO	44 h	Balloon angioplasty; intra-arterial thrombolysis	12 h later: oriented, normal speech, mild hemiparesis (4/5). At 1, 3 and 9-month follow-up, normal

the main reason for the delay in diagnosis was the initial misleading signs and symptoms of the patients. In our case the onset of the syndrome was featured by two “classic” pitfalls: hemiparesis and convulsions. Both of them were mistakenly interpreted as cortical in origin. This is a common phenomenon both in children and adults with ABAO [14,16,17].

The outcome of the reviewed cases with locked-in syndrome with ABAO that received conservative treatment was grim (Table 1). Most of the children remained locked-in or had severe neurological deficit [4,18–23]. In comparison, the 5 children that received endovascular therapy, had a better outcome [5,12–15]. One of them remained locked-in (he received intra-arterial t-PA alone) [14], 2 of them had minimal deficit [5,15] and the other 2 were completely normal after different periods [12,13]. Our patient had the faster recovery: mild hemiparesis (4/5) after 10 days, normal at 1-month follow up.

4. Conclusions

The clinical diagnosis of ABAO in children is often delayed. The reasons for this are the rarity of this condition and the vague, non-specific presenting symptoms. Intra-

arterial thrombolysis combined with cerebral angioplasty, can successfully restore the patency of the basilar artery and the neurologic deficit of children with ABAO, even after a considerable delay. To our knowledge, this is the first pediatric report of complete clinical recovery after delayed endovascular recanalization of BA.

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