Dural Carotid Cavernous Sinus Fistula: Ocular Characteristics, Endovascular Management and Clinical Outcome

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Objective: *To describe the ocular findings, endovascular treatment, and clinical outcome in patients with dural carotid cavernous sinus fistula (CCF).*
Material and Method: *A retrospective evaluation of 80 consecutive patients who underwent examination and treatment for dural CCF between January 1997 and December 2004 was performed.*
Results: *Fifty females and 30 males, with an average age of 49 years (from 6 -80 years) participated in this study. All patients had more than one clinical signs and symptoms including proptosis (84%), arterialization of conjunctival vein (93%), chemosis (42%), cranial nerve palsy (52%), elevated intraocular pressure (51%), and optic neuropathy (13%). Diminished vision was found in 43% of the patients. The degree of visual deficit ranged from 20/40 to no light perception. After angiographic evaluation, patients were classified to CCF Barrow's type B 14%, type C 15%, and type D 71%. Endovascular treatment by transvenous and/or transarterial embolization was performed in 60 patients (75%). Carotid-angular compression therapy was solely performed in 19 patients (24%) and was used as an adjunct to endovascular treatment in 30 patients (38%).The follow-up period ranged from 6 to 94 months. Clinical cure was achieved in 41 patients (51%) and improvement in 30 patients (38%). Anatomical cure was demonstrated by angiogram in 50 patients (63%). Intra-operative complications were found in three patients including ophthalmic artery occlusion and cerebral infarction. Eight patients experienced transient aggravation of symptoms including increased proptosis, elevation of intraocular pressure, choroidal detachment that required suprachoroidal drainage, and venous stasis retinopathy. Ophthalmic vein thrombosis resulting in central retinal vein occlusion was developed in three patients and finally caused severe visual deficit. There was no operative mortality.*
Conclusion: *Selective management with endovascular therapy and manual compression are the effective treatment for dural CCF. However, sight-threatening complications can develop after therapy due to progressive ophthalmic vein thrombosis and should be carefully monitored.*

Keywods: *Carotid cavernous sinus fistula, Endovascular, Outcome*

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Dural carotid cavernous sinus fistula (CCF) is an abnormal communication between the meningeal branches of the internal and/or external carotid artery and the cavernous sinus (CS). It most commonly occurs spontaneously in elderly women, without a definable precipitating factor. The clinical presentation of dural CCF depends on the pathological anatomy of the fistula and venous drainage pattern. Reversal of blood flow in the ophthalmic vein will result in various ophthalmic and orbital symptoms whereas leptomeningeal or cortical drainage will correlate with neurological dysfunction or intracranial hemorrhage.

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For, many years, dural CCFs have been almost exclusively treated by endovascular procedures with transvenous or transarterial embolization. However, the lesions can be treated conservatively in selected cases due to a high incidence (33-50%) of spontaneous regression[1]. Manual compression of the carotid artery has been shown to be effective in some patients[2].

The objective of the present study was to analyze the ocular characteristics, angiographic findings, management, and treatment outcome in Thai patients with dural CCF over an 8-year period.

Material and Method

From January 1997 to December 2004, 80 patients with angiographically confirmed of dural CCF were included in the present study. A retrospective analysis and review was undertaken from the patient's charts. The present study was approved by the Ramathibodi Hospital Ethics Review Board.

Bilateral selective internal carotid artery (ICA), external carotid artery (ECA) angiography and vertebral artery angiography were performed for all patients, for assessment of the feeding arteries, size, sites, and venous drainage pattern of the fistula. According to angiographical findings, Each patient was classified into one of three types of dural CCF, with Barrow type B is an indirect fistula between the CS and dural branches of ICA; Barrow type C is an indirect fistula between the CS and the dural branches of ECA, and Barrow type D is an indirect fistula between the CS and dural branches of both ICA and ECA.

Embolization was performed with single or multiple occlusive agents including liquid adhesive agents, fibered platimun coils, gel foam, or balloon using real-time digital subtraction fluoroscopic mapping. Angiography was done immediately after the completion of the procedure to check for occlusion of the fistula. The techniques of transvenous and transarterial embolization were previously described[3].

Conservative management with carotid artery - angular vein compression was recommended in patients who had single anterior drainage via ophthalmic vein, no cortical drainage and no evidence of cervical carotid atherosclerosis. The patients were instructed to compress the lesion-side carotid artery using their contra-lateral hands and compress the superomedial part of eyelid using their ipsilateral fingers for a period of 15 minutes three times a day.

The patients were followed-up clinically after several days, 1-month, 3-months, 6-months, and yearly intervals. Follow up angiogram was done in each patient to demonstrate complete resolution or residual fistula. The authors defined clinical cure as "free of all the clinical signs and symptoms attributed to the fistula", improved as "symptoms significantly diminished after treatment", stationary as "no change of symptoms", and aggravated as "new symptoms occurred or old symptoms progressed". An anatomical cure was defined by the absence of angiographic evidence of CCF. All data were summarized in the frequency table and bar diagram with number of patients and percentage.

Results

During this 8 years study, 80 patients underwent angiographic evaluation and endovascular procedure for treatment of dural CCF. Of these, there were 50 females and 30 males. The patients' ages ranged from 6 to 80 years, with a median of 51 years (Fig. 1). The follow up periods ranged from 6 to 94 months.

All patients had more than one clinical symptom. The common symptoms were pulsatile tinnitus, blurred vision, and diplopia. The neuro-ophthalmological signs (Fig. 2) were arterialization of conjunctival vein in 93%, proptosis in 84%, chemosis in 42%, bruit in 28% and ophthalmoparesis in 52% (73% abducens, 5% oculomotor, 22% multiple). Diminished visual acuity was found in 43% of the patients. The degree of visual deficit ranged from 20/40 to no light perception. Optic neuropathy, which was diagnosed by the presence of the following findings: decreased visual acuity, abnormal visual field, color vision deficit and positive afferent pupillary defect was found in 13% of patients and elevated intraocular pressure (more than 22 mmHg) was found in 51%. Eight patients had mild degree of

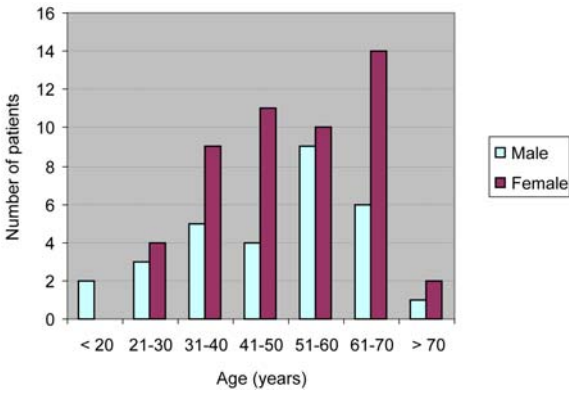

Fig. 1 Distribution of age and sex in 80 patients with dural CCF

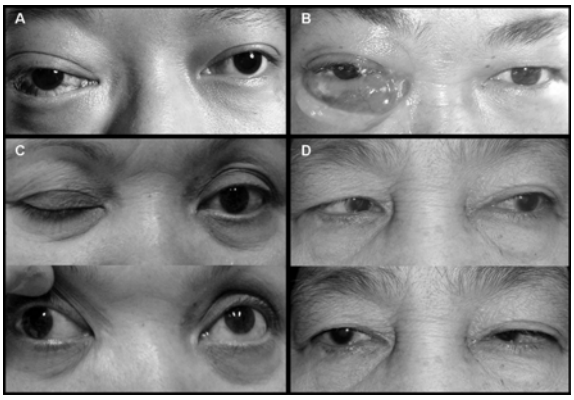

Fig. 2 Different neuro-ophthalmological presentation of patients with dural CCF. (A) Conjunctival injection and proptosis of right eye. (B) Proptosis and severe chemosis of right eye. (C) Isolated right third nerve palsy without orbital signs and symptoms (white-eye syndrome) in a patient with posteriorly draining dural CCF. (D) Bilateral sixth nerve paresis in a patient with bilateral dural CCF

venous stasis retinopathy. No patient in the present series presented with intracranial hemorrhage.

The site of the fistula was on the right in 51%, on the left in 31%, and bilateral in 18%. In the present series, 11 patients (14%) were type B, 12 patients (15%) were type C, and 57 patients (71%) were type D. Venous drainage involved superior ophthalmic vein (SOV) in 85% of patients, inferior ophthalmic vein (IOV) in 7.5%, superior petrosal sinus (SPS) in 20%, inferior petrosal sinus (IPS) in 31%, and contralateral cavernous sinus (via intercavernous sinus) in 9%. Cortical venous drainage was present in 12 patients (15%).

Treatment and outcome of the patients are shown in Table 1. Carotid-angular compression was performed solely in 19 patients (24%). Sixty-eight percent (13/19) of the patients had type-D CCF and 32% (6/19) had type-B CCF. Follow-up angiography was obtained in 13 patients. Of these patients, anatomical cure was demonstrated in eight patients and minimal residual shunt in four patients. Seven patients were cured with respect to clinical symptoms. Ten patients exhibited improvement; most of these cases had residual mild proptosis and arterialization of conjunctival vein. One patient had stationary symptoms and no change of fistula demonstrated by angiography. Subsequently attempted transvenous embolization failed in this patient. One patient had aggravated clinical symptoms.

Endovascular treatment was performed in 60 patients (75%). Transarterial embolization was performed in 40 cases. Transvenous embolization was performed in 34 cases. Fourteen patients were embolized transarterially prior to transvenous embolization. Embolization was performed using a combination of liquid adhesive agents (52%), platinum coils (43%), gel foam (23%), or balloon (5%). Seventeen patients required two or more endovascular treatments to achieve good clinical outcome. Compression therapy

Table 1. Treatment outcome of 79 patients* with dural CCFs: comparison between manual compression and endovascular therapy

	Manual compression therapy	Endovascular therapy
Number of patients	19 (24%)	60 (75%)
Type of CCF	32% type-B, and 68% type-D	7% type-B, 20% type-C, and 73% type-D
Clinical outcome		
Clinical cure	7 (37%)	34 (57%)
Improved	10 (53%)	20 (33%)
Stationary	1 (5%)	3 (5%)***
Aggravated	1 (5%)	3 (5%)
Follow-up angiography		
Number of patients	13	54
Anatomical cure	8 (62%)	42 (78%)
Minimal residual shunt	4 (31%)	12 (22%)
No change of fistula	1 (7%)**	-

* There was a patient who had no treatment and developed spontaneous thrombosis of dural CCF
** Subsequently attempted transvenous embolization was failed in this patient
*** 3 patients with stationary symptoms were reluctant to undergo further aggressive treatment

was used as an adjunct to endovascular treatment in 30 patients. The clinical symptoms had completely disappeared in 34 patients (57%), improved in 20 patients (33%), were stationary in three patients (5%) and were aggravated in three patients (5%). The three patients with stationary symptoms were reluctant to undergo further aggressive treatment. Anatomical cure was demonstrated in 42 patients. Spontaneous thrombosis with resolution of symptoms was developed in one patient who was reluctant to have endovascular treatment and had no therapeutic intervention.

There were six symptomatic complications including two cases of cerebral infarction and four cases of worsening of visual acuity. A patient had a reflux of glue droplets into the petrosquamal branch of the internal maxillary artery resulting in transient minimal facial palsy. Glue particles escaped via the fistula into the middle cerebral artery in another patient causing transient hemiparesis. In four patients with decreased vision, one patient had a reflux of glue into the ophthalmic artery resulting in complete blindness from ophthalmic artery occlusion. Three patients had central retinal vein occlusion that developed after treatment of fistulas. All of these patients finally had severe visual deficit. There were eight patients experiencing transient aggravation of symptoms after treatment including increased proptosis, elevation of intraocular pressure, worsening of ophthalmoparesis, choroidal detachment, which required suprachoroidal drainage and venous stasis retinopathy. There was no operative or perioperative mortality.

Discussion

Intracranial dural arteriovenous fistulas account for 10-15% of all intracranial arteriovenous shunt[4]. The commonest sites of involvement are the transverse sinus and the cavernous sinus[5]. Dural CCF is acquired lesion, thought to originate from an insult to a dural venous sinus that stimulates an inflammatory response with subsequent neovascularization, angiogenesis, and development of a pathological shunt at the arteriolar level[6]. The most common presentation is in perimenopausal women and is least common in men (10-27%)[7,8] suggesting an underlying hormonal influence. However, in contrast to previous literature, the present study found a higher incidence of dural CCF in men (40%). The authors also found that male patients had their onset of dural CCF at a significantly younger age than women.

Dural CCF produces symptoms based on the rate of flow and pattern of venous drainage. Patients with anterior drainage usually present with ophthalmic signs and symptoms including proptosis, pain, chemosis, reduced visual acuity and ophthalmoparesis. However, in some patients with posterior drainage, the ophthalmic presentation is less obvious and the clinical diagnosis is often difficult (so called white-eye syndrome). These patients have a high risk of intracerebral hemorrhage, which may be fatal.

Proptosis, arterialization of conjunctival vessels, and chemosis were the most common findings in the present study. Unlike direct CCF, the degree of proptosis in dural CCF is milder and usually does not lead to sight-threatening complications such as exposure keratitis or corneal ulceration. Eye movement disorders were found in about half of the patients. These abnormalities may result from cranial nerve palsies or congestion and hypoxia of the eye muscles itself. The sixth nerve was the most commonly affected, followed by multiple ocular motor nerve involvement. There was a patient who presented with isolated third nerve paresis with pupillary involvement and orbital pain, a presentation that initially suggested an intracranial aneurysm. The correct diagnosis of dural CCF in this case was not evident until cerebral angiography was performed.

Decreased visual acuity in patients with dural CCF may be caused by ischemic optic neuropathy, chorioretinal dysfunction, or glaucoma. Increased intraocular pressure resulting from raised episcleral venous pressure was not an uncommon finding. Fifty-one percent of patients in the present study were diagnosed with secondary open angle glaucoma. Among these cases, IOP was normalized after closure of fistula in 72% (40/55) of cases and there were 15 patients who still need antiglaucoma medication for controlling their IOPs. No patient in the present study underwent a filtration surgery.

Carotid artery compression has been reported as an effective treatment of dural CCF[2]. By compressing the carotid artery, antegrade flow is diminished and venous pressure elevated promoting thrombosis of the fistula[6]. This technique can be an adjunct to achieve complete obliteration of the fistulae in patients with partial thrombosis or subtotal angiographic cure. The present study showed that good clinical outcome was obtained after perform manual compression in 89% of selected cases. Liu et al[1] recommend conservative treatment as the first choice for the patient with type-B CCF and reserved embolization for cases with aggressive signs such as cortical venous drainage, malignant proptosis, and increased intraocular pressure.

Endovascular management is primarily the treatment in dural CCF. Rarely, direct surgery on the cavernous sinus is required. Recently, the transvenous or internal carotid approach has become the best therapeutic option for patients with symptomatic dural CCF. The transvenous route is preferred to the transarterial route, due to higher clinical and anatomical cure rates and a lower incidence of complications[3,9,10]. In Ramathibodi Hospital, transvenous embolization has become the treatment of choice in most cases of dural CCF since 2004. Transvenous access of the inferior petrosal sinus is readily available even if thrombosed and is frequently used (Fig. 3).Transarterial embolization is used only for some cases of type-C fistula or as additional therapy after transvenous embolization.

The occurrence of spontaneous thrombosis of the dural CCF with clinical and angiographic cure has been well documented[1]. In the present study, the authors cannot corroborate the frequent rate of spontaneous resolution and documented this phenomenon in only one patient after diagnostic angiography. However, the same mechanisms that result in spontaneous closure of fistula may play some role in several patients who progressed to complete fistula occlusion following subtotal endovascular treatment and in patients who were successfully treated with manual compression therapy.

Following successful treatment, patients may experience an aggravation of their clinical symptoms a few weeks before their symptoms totally disappear. This transient worsening may be caused by the inflammatory effects of thrombosis in the highly enervated cavernous sinus or propagation of thrombus throughout the cavernous sinus extending into the superior

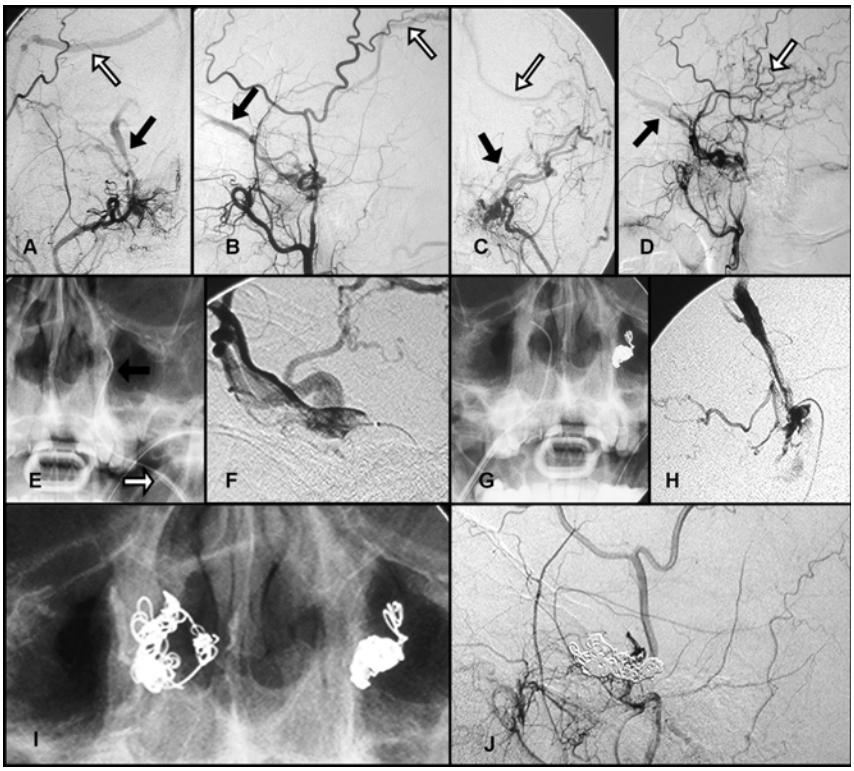

Fig. 3 A 52-year-old man presented with bilateral proptosis. (A) and (B), right ECA angiogram (anteroposterior and lateral projections), showing a right dural CCF with venous drainage into the right SOV (black arrow) and right superficial Sylvian vein (white arrow). The right IPS is occluded. (C) and (D), left ECA angiogram, showing a left dural CCF with venous drainage into left SOV (black arrow) and left middle cerebral vein (white arrow). The left IPS is occluded. (E) and (F) transvenous approach through occluded left IPS using 038 guidewire (black arrow) and 6 Fr. guiding catheter (white arrow) with mechanical re-opening technique, followed by coil embolization. (G) and (H) transvenous coil embolization via right IPS using the same technique. (I) Water's view plain film after successful embolization into bilateral cavernous sinus. (J) control angiogram shows total occlusion of dural CCFs

ophthalmic vein[7]. Increasing venous pressure resulting from the process of thrombosis in the superior ophthalmic vein may be also the cause of this paradoxical phenomenon[11]. Spontaneous resolution usually occurs over time but a brief course of steroid may help to reduce inflammation associated with acute thrombosis. Three patients in the present study developed central retinal vein occlusion after treatment of fistulas and finally had severe visual deficit. This serious complication might have resulted from permanent ophthalmic vein thrombosis.

In conclusion, selective management with endovascular therapy and manual compression are the effective treatment for dural CCF. However, sight threatening complications can develop after therapy due to progressive ophthalmic vein thrombosis and should be carefully monitored. Future advances in endovascular equipment will expand on the current applications and further minimize procedure-related complications.

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การศึกษาลักษณะทางตาและผลลัพธ์ทางคลินิก ในผู้ป่วยที่มีทางทะลุระหว่างหลอดเลือดแดงคาโรติดและโพรงเลือดคาเวอร์นัสชนิดดูรา ซึ่งได้รับการรักษาโดยวิธีใส่สายสวนผ่านทางหลอดเลือด

พิศิษฐ์ ปรีชาวัฒน์, พิสัณห์ นามเกิด, ปกรณ์ เจียระคงมั่น, อนุชิต ปุญญทลังค์, ศิรินทรา พงษ์เพ็ชร

วัตถุประสงค์: *เพื่อศึกษาลักษณะทางตาและผลลัพธ์ทางคลินิก ในผู้ป่วยที่มีทางทะลุระหว่างหลอดเลือดแดงคาโรติดและโพรงเลือดคาเวอร์นัสชนิดดูรา (dural CCF) ซึ่งได้รับการรักษาโดยวิธีใส่สายสวนผ่านทางหลอดเลือด*

วัสดุและวิธีการ: *ศึกษาข้อมูลย้อนหลังจากเวชระเบียนผู้ป่วยที่ได้รับการวินิจฉัยว่าเป็น dural CCF ตั้งแต่เดือนมกราคม พ.ศ. 2540 จนถึง เดือนธันวาคม พ.ศ. 2547*

ผลการศึกษา: *จำนวนผู้ป่วยทั้งหมด 80 ราย เป็นผู้หญิง 50 ราย และผู้ชาย 30 ราย พบผู้ป่วยได้ตั้งแต่ อายุ 6 ถึง 80 ปี อายุโดยเฉลี่ย 49 ปี อาการและอาการแสดงที่พบ ได้แก่ ตาโปน (84 เปอร์เซ็นต์) หลอดเลือดดำบริเวณเยื่อบุตาขยายตัว (93 เปอร์เซ็นต์) เยื่อบุตาบวมน้ำ (42 เปอร์เซ็นต์) การกลอกตาผิดปกติ จากอัมพาตของเส้นประสาทสมอง (52 เปอร์เซ็นต์) ความดันลูกตาสูง (51 เปอร์เซ็นต์) และความผิดปกติของเส้นประสาทตา (13 เปอร์เซ็นต์) พบว่า 43 เปอร์เซ็นต์ของผู้ป่วยมีระดับการมองเห็นที่ลดลง ตั้งแต่ 20/40 จนถึง มองไม่เห็นแสง จากการฉีดสีดูหลอดเลือด พบว่า dural CCF เป็น ชนิด B 14 เปอร์เซ็นต์ ชนิด C 15 เปอร์เซ็นต์ และ ชนิด D 71 เปอร์เซ็นต์ ผู้ป่วย 60 ราย (75 เปอร์เซ็นต์) ได้รับการรักษาผ่านทางสายสวนหลอดเลือด การรักษาโดยการกดหลอดเลือดแดงแคโรติดที่คอเพียงวิธีเดียวทำในผู้ป่วย 19 ราย (24 เปอร์เซ็นต์) และทำร่วมกับการรักษาผ่านทางสายสวนหลอดเลือด จำนวน 30 ราย (38 เปอร์เซ็นต์) ระยะเวลาในการติดตามการรักษาตั้งแต่ 6 ถึง 94 เดือน ลักษณะทางคลินิกหายขาดในผู้ป่วยจำนวน 41 ราย (51 เปอร์เซ็นต์) ดีขึ้น 30 ราย (38 เปอร์เซ็นต์) จากการฉีดสีดูหลอดเลือดหลังการรักษา พบว่ารอยรั่วหายสนิทในผู้ป่วย 50 ราย (63 เปอร์เซ็นต์) ภาวะแทรกซ้อนในขณะทำการรักษา พบในผู้ป่วย 3 ราย ซึ่งเกิดการอุดตันของหลอดเลือดแดงออฟทัลมิก หรือ หลอดเลือดแดงในสมอง หลังการรักษา พบว่ามีผู้ป่วย 8 ราย ที่มีอาการบางอย่างกำเริบมากขึ้น เช่น ตาโปนมากขึ้น ความดันตาสูงขึ้น และซั้นคอรอยด์มีการหลุดลอก อย่างไรก็ตามภาวะต่าง ๆ เหล่านี้เกิดขึ้นเพียงชั่วคราว พบว่ามีผู้ป่วย 3 ราย ที่เกิดการอุดตันของหลอดเลือดดำใหญ่ในจอตาหลังการรักษา และระดับการมองเห็นลดลงอย่างมาก*

สรุป: *การเลือกวิธีการรักษาที่เหมาะสม โดยวิธีการรักษาผ่านทางสายสวนหลอดเลือด หรือ การกดหลอดเลือดแดงคาโรติดที่คอ ได้ผลดีในการรักษาผู้ป่วย dural CCF อย่างไรก็ตามควรให้ความระมัดระวังภาวะแทรกซ้อนที่สำคัญที่อาจเกิดขึ้นหลังการรักษาได้แก่ การเกิดการอุดตันของหลอดเลือดดำใหญ่ในจอตา*