Beth Israel Lahey Health
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The Brain Aneurysm Institute

Multidisciplinary Care of Patients with Hemorrhagic and Ischemic Stroke

WINTER 2023

Neurovascular News

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BIDMC Polycystic Kidney Disease program accredited as Center of Excellence

Peter G. Czarnecki, MD, Max Shutran, MD, Philipp Taussky, MD, Christopher S. Ogilvy, MD

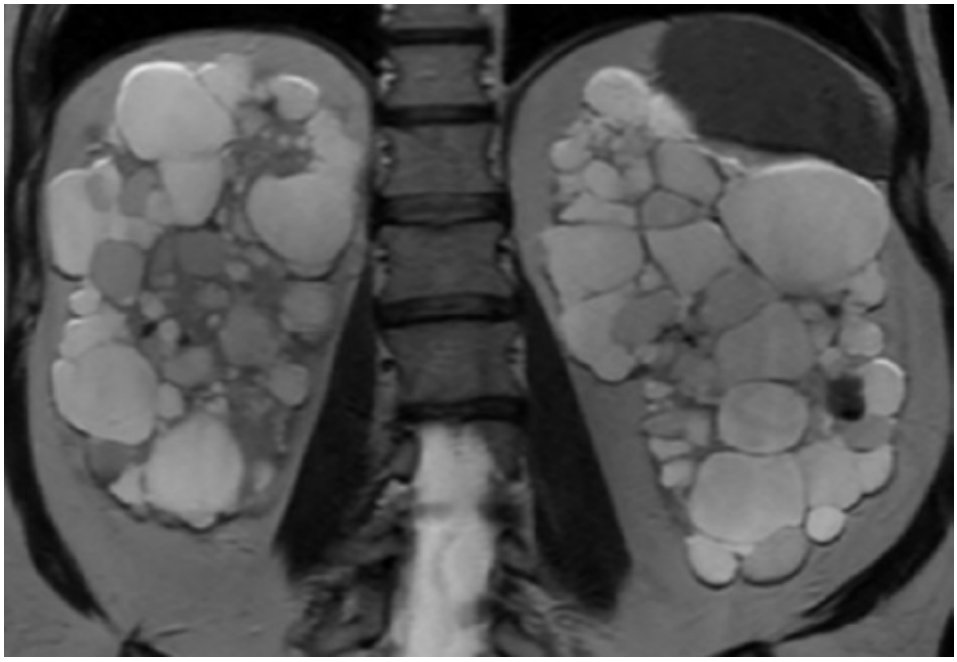
The BIDMC Polycystic Kidney Disease (PKD)- and Kidney Genetics Clinic are proud and pleased to have received recognition by the PKD Foundation as a ***Center of Excellence in PKD care***. Patients with Polycystic Kidney Disease have a much higher risk of developing an intracranial aneurysm and suffer from aneurysm rupture and subarachnoid hemorrhage. Interdisciplinary collaboration in the detection, risk stratification and long-term follow-up is key to successful management. The BIDMC Brain Aneurysm Institute has joined with the PKD program to bring integrated care and excellence to our patients.

PKD epidemiology, clinical presentation and course: With approximately 1 in 400-1000 live births, autosomal-dominant Polycystic Kidney Disease (ADPKD) is one of the most common genetic disorders in mankind[1]. Approximately 600,000 Americans, and about 12 million people worldwide are affected.[2,3] The disease is characterized by the presence and life-long expansion of cysts in both kidneys. These cysts are fluid-filled sacs that occupy progressively more anatomical space as they grow, compressing the residual healthy kidney tissue and impairing overall kidney function (Figure 1). Ultimately, patients suffer from worsening kidney failure, terminating in end-stage kidney disease (ESKD) and the need for dialysis or kidney transplantation. In the most common form of ADPKD, patients reach ESKD at an age between 50 and 60. It is in its nature as a genetic disease that every cell in the patient's body carries the causative gene mutation. While the kidney is the most affected target organ, other tissues often exhibit effects of the disease. Cysts can often be found in the liver and pancreas, and some patients have abnormalities of heart valves and blood vessels. Of all these extrarenal manifestations, the most dangerous is the presence of intracranial aneurysms.

While 1-2% of the general population may carry an intracranial aneurysm, the risk in patients with PKD is 4-5 times higher (4-8%). Patients with PKD who have a family history of intracranial aneurysms have an even higher risk of carrying an aneurysm themselves (approximately 20-25%)[4]. As advanced kidney dysfunction or dialysis treatments in the context of PKD are associated with high blood pressure, many patients with an undiscovered intracranial aneurysm are at risk of a subarachnoid hemorrhage that is often fatal. Given the grave consequences of a subarachnoid hemorrhage, it is of key importance to identify any existing aneurysms in this high-risk population, before they reach a size at which rupture becomes more likely.

PKD patients typically undergo a screening MRA scan as young adults. Once an aneurysm is discovered, it is evaluated for its rupture potential. Criteria include, but are not limited to anatomical location, size and geometry. While small aneurysms can often be monitored with repeat MRA scans, large aneurysms often require open surgery or endovascular therapy to prevent a future rupture event. In addition, aneurysms in patients with ADPKD can rupture at a smaller size compared to other patients with aneurysms.

Figure 1: MRI scan of t he kidneys of a patient with advanced polycystic kidney disease



Interdisciplinary approach to long-term management: Established by the PKD Foundation, the PKD Center of Excellence is an elite designation for clinics helping those affected by PKD better manage their disease, maintain and improve their quality of life[5]. Centers of Excellence provide comprehensive, multidisciplinary clinical services for families affected by ADPKD. Services are coordinated through the center to define an individual's clinical care needs. At BIDMC, the PKD and Kidney Genetics Clinic serves as the core site, evaluating new patients with PKD, providing second-opinion consultations and longitudinally managing local patients. Multidisciplinary collaborations with the Brain Aneurysm Institute, the Divisions of Kidney Transplantation Medicine and Surgery, and Hepatology are the cornerstones of the specialist provider network. While improving lives today, the Centers of Excellence will drive research advancements and quality care by increasing research opportunities, collaboration, mentorships, communication and education.

A patient with ADPKD and aneurysm: A 61 year old woman with a history of ADPKD had undergone successful kidney transplant several years earlier. Brain MRI studies had disclosed a small left anterior cerebral artery aneurysm that grew slightly over 3 years of follow up at the BIDMC Brain Aneurysm Institute. Interestingly, she also had an asymptomatic occlusion of the middle cerebral artery on the left side. After extensive discussion and review with our multidisciplinary team, she elected to have surgery with a craniotomy to clip the aneurysm. Figure 2A and 2B demonstrate the aneurysm as seen on the cerebral angiogram. Figure 3A and 3B show the intraoperative view of the aneurysm before (3A) and after (3B) clip obliteration.

The patient awoke from surgery without any neurologic deficit and was discharged home in 48 hours. She has continued to make an excellent recovery, now living without the risk of hemorrhage from her aneurysm.

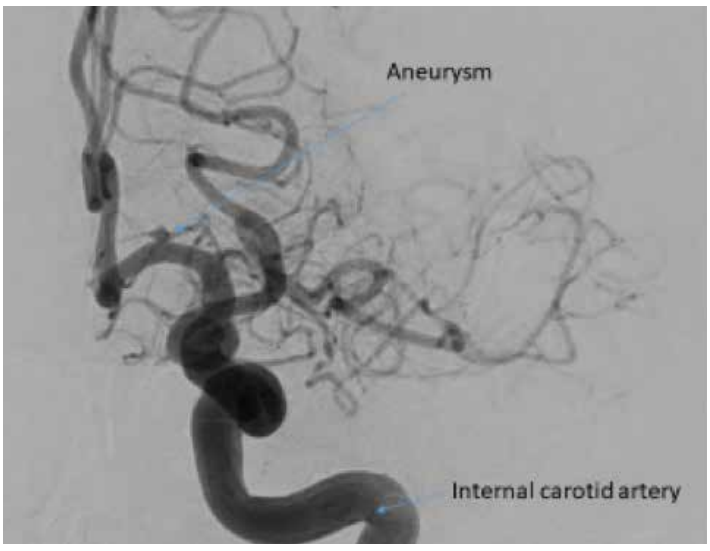


Figure 2A: A-P angiogram demonstrating the A1 segment intracranial aneurysm

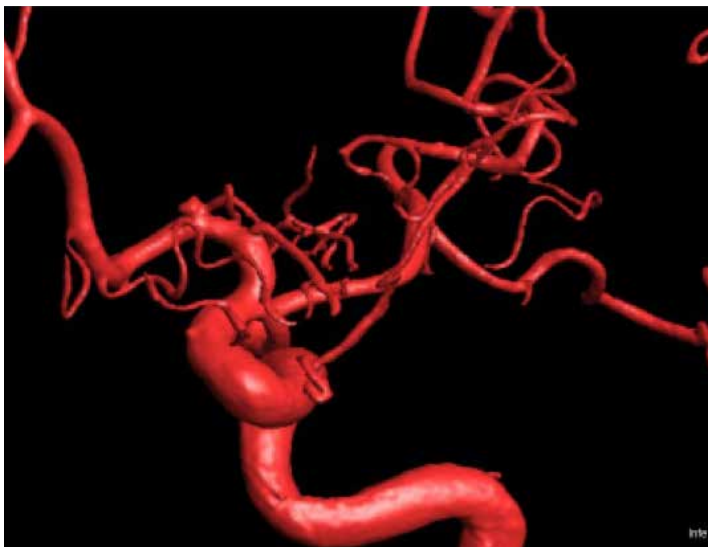


Figure 2B: 3-D cerebral angiogram demonstrating the irregularly shaped A1 segment aneurysm associated with a large perforating artery.

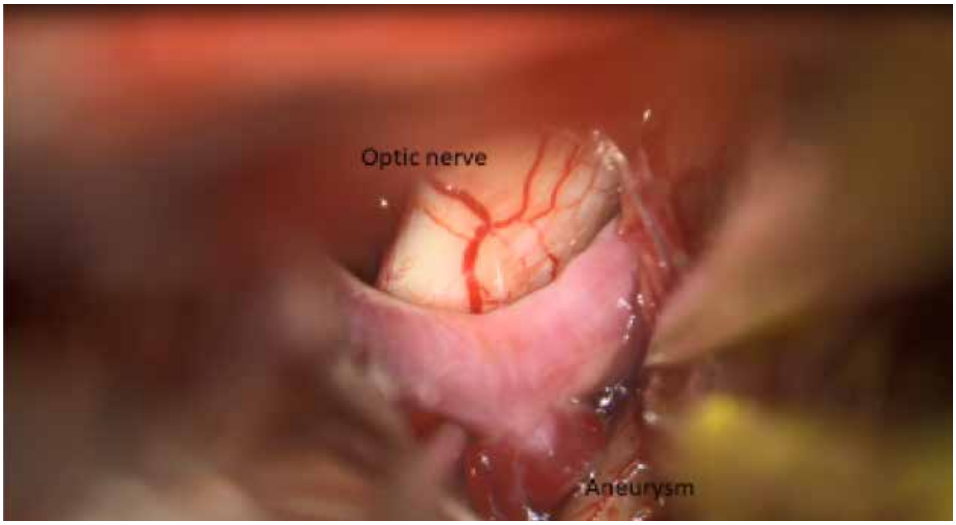


Figure 3A: Intraoperative view showing the small, very fragile, thin walled aneurysm.

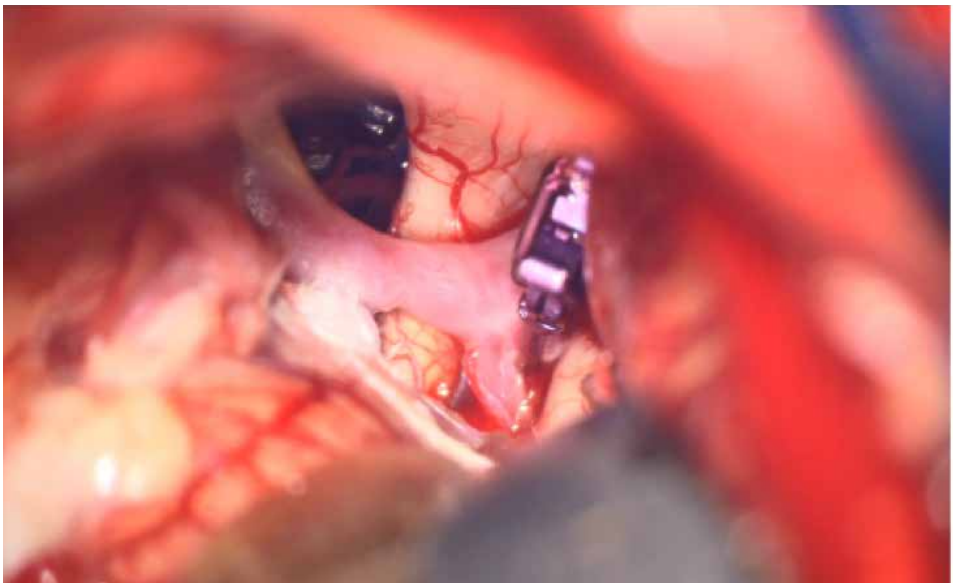


Figure 3B: intraoperative view showing the aneurysm clip in place with good preservation of surrounding normal vessels. The chronically occluded middle cerebral artery can be seen in the lower left of the image.

The future: Many questions in the disease pathogenesis of PKD and brain aneurysms are still unanswered. Through improved access to systematic genetic testing, new PKD genes are being identified, and within the known disease genes, the catalog of novel variants is rapidly growing. It remains unknown if genetic variants in the newly discovered "minor" PKD genes are associated with the same risk for brain aneurysm formation as in classic ADPKD. Similarly, so-called ultra-low-penetrance alleles in the major PKD genes are quite common in the general population, without giving rise to kidney disease. However, if those variants were associated with an increased brain aneurysm risk, this would dramatically change the diagnostic landscape and screening recommendations. The collaboration between the BIDMC Brain Aneurysm Institute and the PKD Clinic will therefore go beyond its clinical role in multidisciplinary patient management, and also explore research opportunities across specialties, addressing unmet needs in our understanding of brain aneurysm formation.

References:

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2. National Institutes of Health Facts Sheet on PKD, https://www.niddk.nih.gov/-/media/Files/Kidney-Disease/PKD_508.pdf
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4. Perrone RD et al., Nat Rev Nephrol 2015
5. The PKD Foundation, https://pkdcure.org/carecenters

A common question about aneurysms: who should be screened?

- Patients with 2 or more first degree relatives with aneurysm or subarachnoid hemorrhage
- Patients with autosomal dominant polycystic kidney disease particularly with family history
- Initial screening in young adulthood: age 20-25 years old-Should repeat screening at 7-10 year intervals
- Reasonable to screen patients with aortic coarctation and primordial dwarfism
- Women who smoke cigarettes

Dual lumen balloon assisted coiling and staged flow diversion for ruptured, wide necked posterior communicating artery aneurysm

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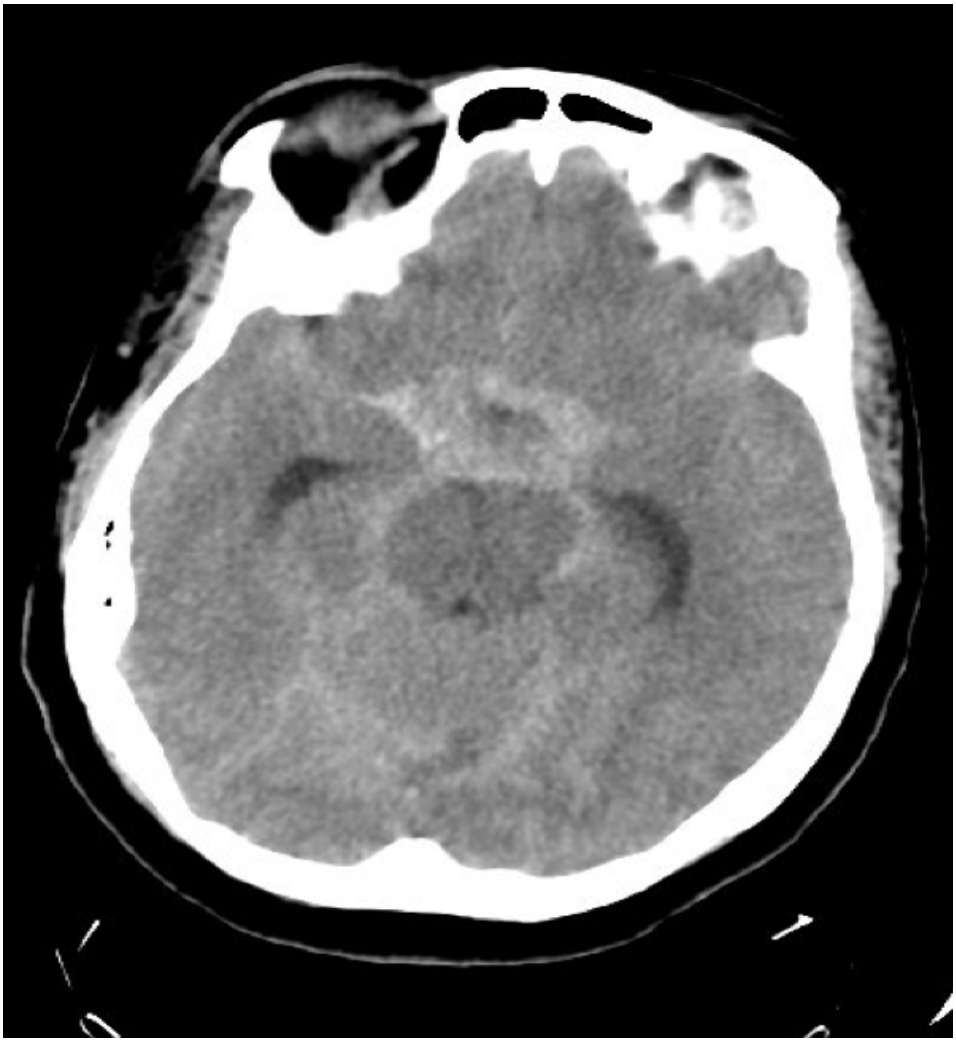


Figure 1: Head CT showing diffuse subarachnoid hemorrhage and hydrocephalus

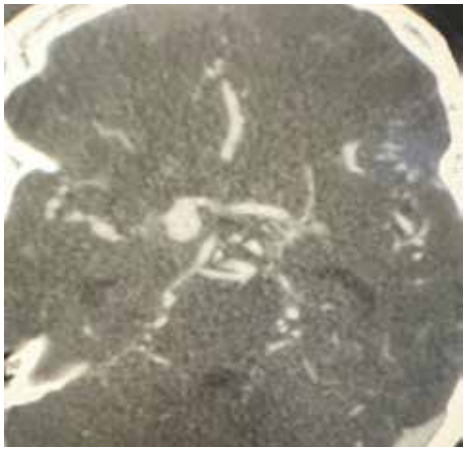


Figure 2A: Axial CTA demonstrating wide neck right posterior communicating artery aneurysm

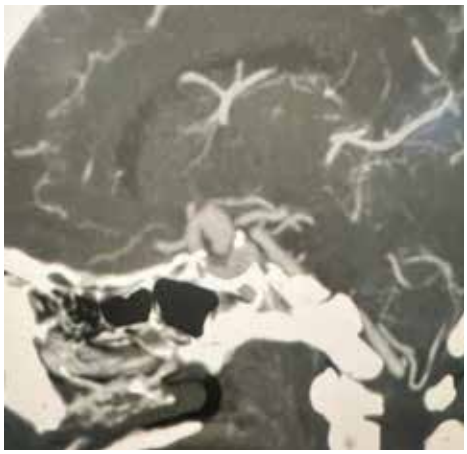


Figure 2B: Sagittal CTA demonstrating wide neck right posterior communicating artery aneurysm

Endovascular treatment of acutely ruptured wide-necked aneurysms presents well-known challenges due to the desire to avoid intracranial stenting with dual antiplatelet therapy requirements. Balloon assisted coiling (BAC) has been well described for this purpose, most commonly using a two-microcatheter technique with a balloon microcatheter protecting the aneurysm neck and a coiling microcatheter used to embolize the aneurysm[1,2]. However, the availability of advanced dual-lumen balloon microcatheters with coiling markers allows for the use of a single microcatheter technique in select instances[3]. We present the case of a 65-year-old female who presented as a transfer to the BIDMC Brain Aneurysm Institute from an outside institution after developing the worst headache of her life. Prior to transfer from the outside institution, she was intubated due to increased lethargy and concerns for airway protection. Upon arrival, her neurologic examination had deteriorated and she was only able to weakly withdraw her extremities and without eye opening to painful stimuli. Head CT showed thick subarachnoid hemorrhage with the presence of hydrocephalus. (Figure 1) CTA head demonstrated a wide necked posterior communicating artery aneurysm incorporating a large posterior communicating artery at the neck of the aneurysm (Figure 2A/B). An external ventricular drain was emergently placed at the bedside which was shown to be under high pressure. The patient was taken to the endovascular suite for planned subtotal coiling of the aneurysm followed by planned delayed treatment with flow diversion while the patient was hospitalized. Cerebral angiogram demonstrated a

9.09mm x 7.10mm right posterior communicating artery aneurysm with a 5.70mm neck. (Figure 3A/B/C) Due to the presence of a wide necked aneurysm with the large posterior communicating artery origin within the neck of the aneurysm, the decision was made to protect the dome of the aneurysm and subsequent rupture site. The aneurysm dome had sufficient height to allow balloon assisted coiling using a single balloon microcatheter, which allowed for several advantages in treatment. First, the balloon could be inflated within the parent vessel, internal carotid artery, in order to provide cessation of anterograde blood flow in the case of intraprocedural aneurysm rupture. Second, with the balloon inflated both the posterior communicating artery as well as the internal carotid artery would be protected against coil prolapse out of the aneurysm. Third, the dual lumen balloon microcatheter allowed for balloon protection along the same microcatheter used for coiling without the need for two separate microcatheters. The dual lumen balloon microcatheter was inflated for the initial coiling and then subsequently deflated in between each coil in order to check for coil prolapse. (Figure 4) The final angiogram of the balloon assisted coiling procedure showed dense packing within the neck of the aneurysm without compromise of the internal carotid artery or posterior communicating artery. (Figure 5A/B) The patient was then managed in the neuro intensive care unit with significant improvement in her neurologic examination.

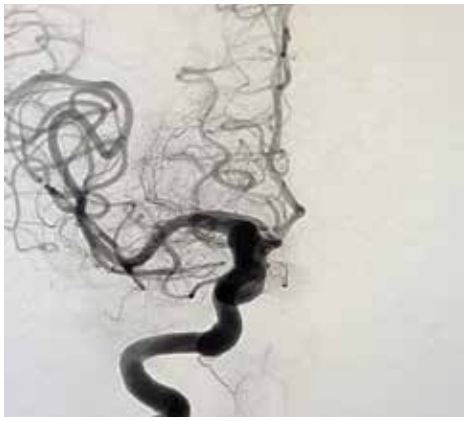


Figure 3A: Anterior posterior cerebral angiogram

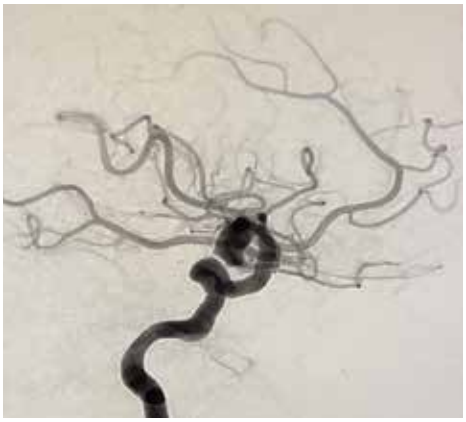


Figure 3B: Lateral cerebral angiogram wide necked right posterior communicating artery aneurysm

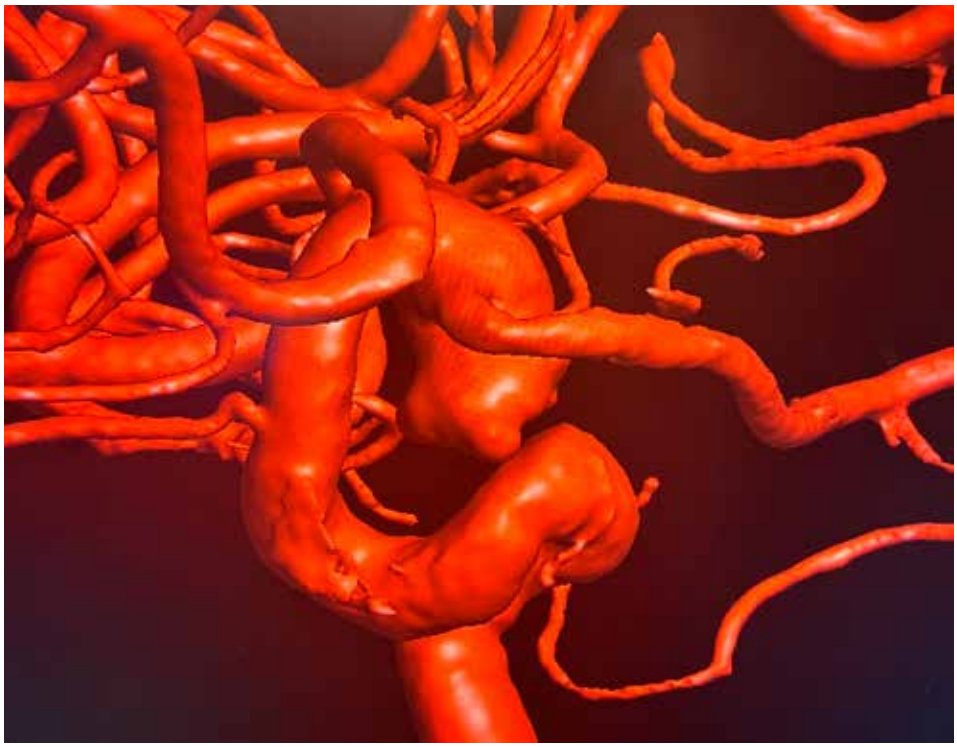


Figure 3C: 3D Reconstruction of wide necked right posterior communicating artery aneurysm incorporating right posterior communicating artery aneurysm

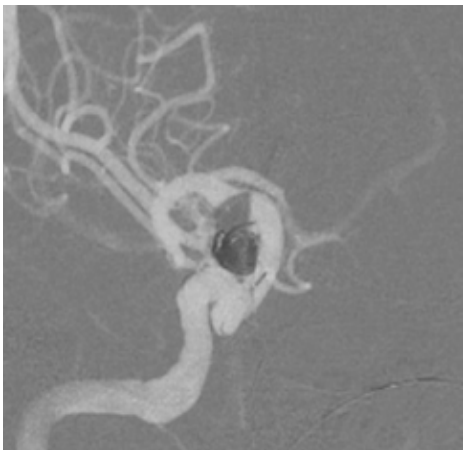


Figure 4: Inflation of dual lumen balloon catheter across the neck of the aneurysm with the tip of the catheter within the aneurysm to allow for coiling

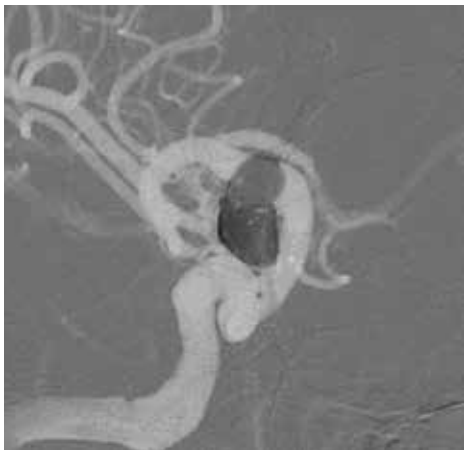


Figure 5A: Angiogram demonstrating inflation of balloon for dense packing of aneurysm dome

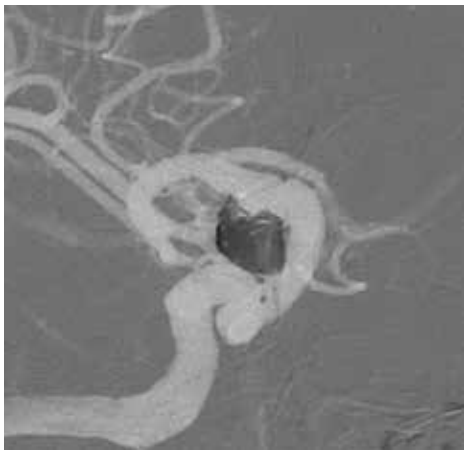


Figure 5B: Angiogram demonstrating deflation of the balloon with no evidence of coil prolapse

Figure 6A: Unsubtracted angiogram post pipeline deployment across the neck of the posterior communicating artery aneurysm

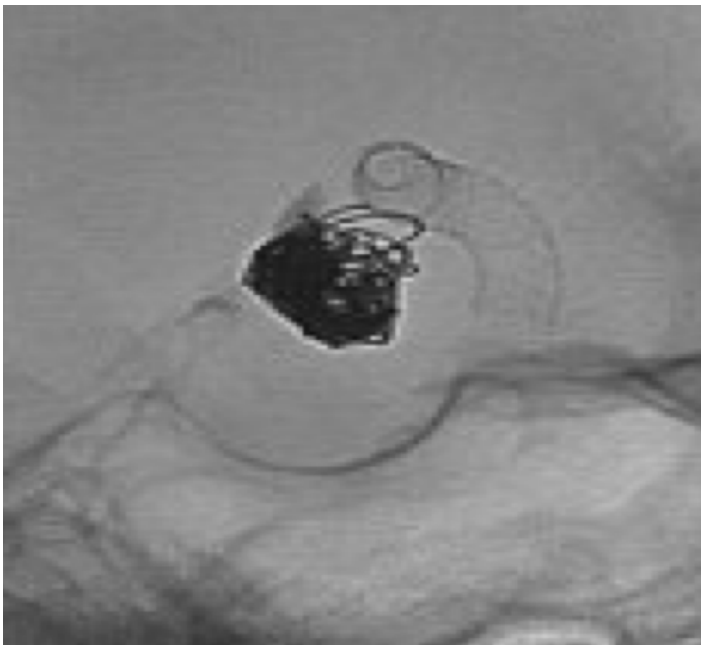
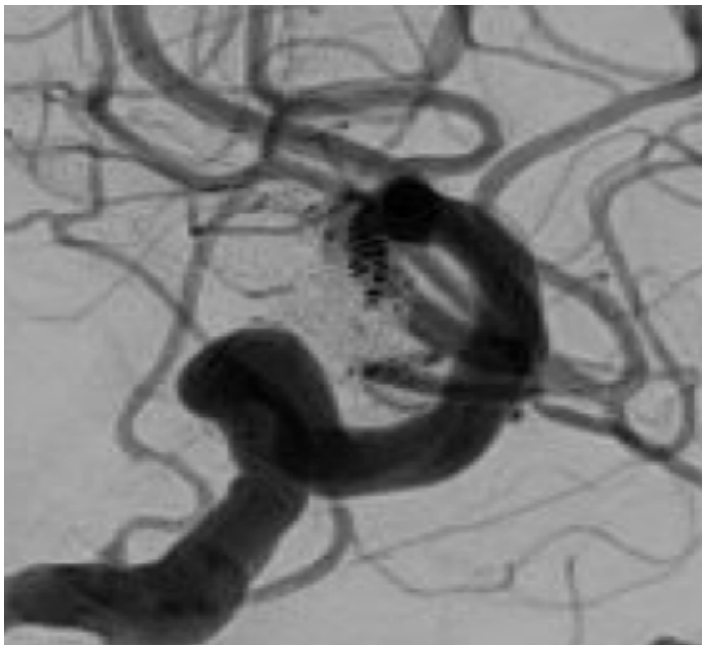


Figure 6B: Lateral angiogram post pipeline deployment across the neck of the posterior communicating artery aneurysm



She was able to be successfully extubated and the external ventricular drain weaned and removed. 3 weeks after her initial presentation, she was started on dual antiplatelet therapy with plan for flow diversion treatment of the residual aneurysm. She underwent pipeline embolization across the neck of the posterior communicating artery aneurysm with aneurysmal stasis post deployment and no evidence of thromboembolic complications. (Figure 6A/B) The next day she was discharged home in neurologically intact condition. This case shows that sub-total aneurysm coiling with delayed flow diversion is a pragmatic strategy in the treatment of wide necked ruptured aneurysms[4], and the use of a single balloon microcatheter for BAC can be useful in certain situations. At the BIDMC Brain Aneurysm Institute we have utilized this novel technique in several patients with good success.

References:

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Current management of small intracranial aneurysms

Christopher S. Ogilvy, MD and Philipp Taussky, MD

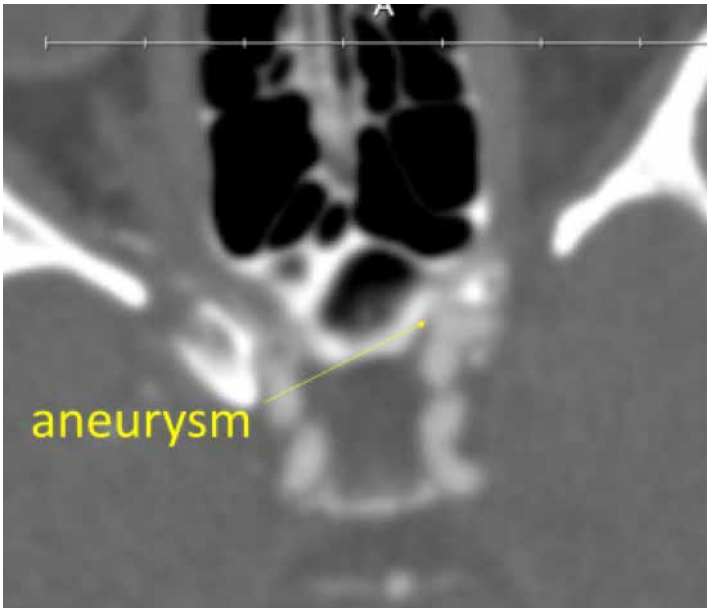
Small intracranial aneurysms are usually defined as lesions less than 10 mm in diameter. In 1998 the international study of unruptured intracranial aneurysms was published in the New England Journal of Medicine[1]. This was a large (60 international centers) nonrandomized trial that had a prospective component and a retrospective component. Patients treated were followed prospectively and the outcomes from surgery were found to have a higher incidence of physical and cognitive outcomes than had previously been reported in retrospective and single center studies. In the prospective arm of the study patients with unruptured lesions were followed and those with aneurysms smaller than 7 mm were found to have a rupture rate which was much lower than had previously been published and on the order of 0.5 %/year. Given these results, it might be anticipated that the number of unruptured aneurysms treated in United States would diminish. However the opposite was observed and more unruptured aneurysms were treated over the subsequent years after the study was released[2].

This could be explained by several factors. For one, image techniques were improving rapidly such that aneurysms are now detectable down to sizes of 1 and 2 mm. In addition, endovascular techniques to treat aneurysms were rapidly evolving and producing outcomes for certain lesions that were safer than "open" surgery.

It was published in a previous addition of this newsletter we are indeed seeing smaller aneurysms treated over the last 20 years. Many of the unruptured aneurysms currently

Figure 1: 83 year old woman with worsening memory where a CTA disclosed a 4 mm internal carotid artery aneurysm. The lesion is incidental and given her age and neurologic condition, treatment would not be considered.

Figure 2: A 48 year old woman with a 40 pack year smoking history and a strong family history of intracranial aneurysms was found to have a 4 mm internal carotid artery aneurysm. Given her age and history should treatment be considered?



detected are smaller than 10 mm particularly those that are incidental. The topic of whether or not to treat a smaller unruptured aneurysm is also complicated by the fact that the majority of the aneurysms that rupture are small. A recent study documented data over 20 years demonstrating that currently 50% of ruptured aneurysms are smaller than 5 mm and 84% are smaller than 9 mm[3]. Thus when a patient with an unruptured aneurysm is encountered in the office they are given information that smaller unruptured aneurysms are less likely to bleed yet when aneurysms bleed they are likely to be small! There has yet to be an adequate full explanation for these data sets.

There do seem to be populations of patients where aneurysms can rupture at a smaller size. There is data to support this concept for patients with autosomal dominant polycystic kidney disease as well as those with a family history of aneurysms and patients with a history of cigarette smoking may also fall into this category. All of these factors fallen to patient related risk factors that are utilized in the decision making of whether or not to treat a small unruptured aneurysm.

Consider two separate patients: Figure 1 shows the CT angiogram from an 83-year-old woman who had the scan done as a screening process for worsening memory. She was found to have a 4 mm internal carotid artery aneurysm. The lesion is completely incidental and given her age with a lower life horizon, treatment would not be considered. In addition, treatment related risks in an older patient are certainly higher than younger individuals. Figure 2 shows the CT angiogram of a 48-year-old woman with a very strong family history of aneurysms where to direct relatives have died of aneurysmal hemorrhage. She herself had a 40-pack-year smoking history. Screening CT angiogram demonstrates a 4 mm internal carotid artery aneurysm. Given her constellation of risk factors and the fact she is young a discussion can be undertaken as to whether this lesion should be treated. Current treatment techniques would predict the risk of the treatment summary on the order of 2.5 to 3% chance of a neurologic or systemic problem.

In addition to the tangible risk factors associated with an aneurysm in predicting risk of rupture and treatment related morbidity based on patient's specific factors there is also the issue of patient anxiety. Intracranial aneurysms can generate significant anxiety once the diagnosis is made. Patients search the Internet and find many stories and testimonial's of patients who have suffered subarachnoid hemorrhage or had succumbed to a ruptured aneurysm.

Based on the constellation of aneurysm specific factors and patient-specific factors extensive discussions are undertaken at the BIDMC Brain Aneurysm Institute with patients and their families as to whether or not a small unruptured aneurysm should be treated. Very often much more time spent counseling patient's why they do not need treatment rather than why they do need treatment. Patient's specific natural history risk profiles can be generated as can treatment related risks for each patient. In the future it is entirely possible that artificial intelligence will help with this process in terms of the radiologic monitoring of these patients as well as incorporation of treatment related risks into the analysis.

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NEWS

The BIDMC Brain Aneurysm Institute is proud to announce the appointment of Max Shutran, MD to the team.

He recently completed his fellowship in Endovascular and Operative Neurovascular Surgery under the leadership of Christopher S. Ogilvy, MD and Philipp Taussky, MD. He received his medical degree from Tufts University School of Medicine and completed a 7-year Neurosurgery residency from the same institution. As a dual trained neurosurgeon, he will be treating patients for both microsurgical and minimally invasive endovascular treatment for vascular disease.

He is involved with various research studies at BIDMC Brain Aneurysm Institute. With the team, he conducts clinical and basic research, as well as research in technology development, in the areas related to neurovascular conditions.

He is dedicated to providing comprehensive, well-coordinated and compassionate care that offers the best possible outcome for the patient and family.

He will be running the Boston Marathon on Monday, Apr 17, 2023 for the Brain Aneurysm Foundation, a 501-C3 nonprofit that is recognized as a leader in brain aneurysm awareness, education, support, advocacy and research funding.

He provides consultation and management for patients with AVMs, aneurysms, cavernous angiomas, meningiomas, Moya-Moya, stroke, carotid disease, and other cerebral problems.

SAVE THE DATE

Ischemic and Hemorrhagic Update: Current Practices and Future Directions

MONDAY, MAY 8, 2023

LOCATION: Omni Parker House Hotel, Boston, MA 02108

This is a unique course focused on recent advances in the field of neurovascular disease including up-to-date theories of carotid disease, cerebral hemorrhage, and brain aneurysms. Topics covered will include assessment, management, and specific issues of carotid disease, cerebral hemorrhage, and brain aneurysms. This will be a hybrid course.

CME credit awarded.