Intracranial aneurysm in tuberous sclerosis complex

Tuberous sclerosis complex (TSC) is a rare multisystem genetic disease. Arterial wall developmental disorders, such as aneurysms, in association with TSC have been well described for extracranial vasculature. The characteristics of intracranial aneurysms (IAs) in TSC have not previously been addressed in the literature. This systematic review was performed to identify and assess the distinct characteristics of IAs in patients with TSC.

METHODS
The authors searched PubMed, Scopus, and Web of Science for publications describing cases of TSC and IA reported before August 7, 2018. They also report 2 cases of IAs in TSC patients treated at their own institution.

RESULTS
Thirty-three TSC patients with a total of 42 IAs were included in this review. Three individuals presented with subarachnoid hemorrhage. The IAs were large or giant in 57.1% and fusiform in 45.2% of the cases. Most of the IAs (61.9%, 26 of 42) originated from the internal carotid artery. There was a higher prevalence of pediatric cases (66.7%) and male patients (63.6%, 21 of 32 individuals with known sex) among the collected series.

TSC patients with IAs are characterized with a higher proportion of large/giant and fusiform IAs and young age, suggesting rapid aneurysmal growth. Furthermore, there is a distinct location pattern of IAs and an inverse sex ratio than in the healthy population. Large population-based patient registers are required to improve the understanding of epidemiology and pathophysiology of IA formation in TSC.  