On-line Table: Demographics, clinical presentation, and neuroimaging findings

Patient	Age (yr)/Sex/ Ethnicity	Main Clinical Findings	Typical *ACTA2* Neurovascular Abnormalities[a]	Bending/Hypoplasia Anterior Corpus Callosum	Abnormal Radial Gyration Frontal Lobes	Cortical Malformations	Absent Anterior Cingulate Gyrus	Twin Peaks Pons	Squeezed Midbrain	Paramedian Indentation of Pontine Surface	Large Territorial Infarctions	Watershed Infarctions
1	23 months/F/Pakistani	PDA, congenital mydriasis, pulmonary hypertension, right hemiparesis	Yes	Yes	Yes	No	Yes	No	No	No	Yes	Yes
2	4/F/white	PDA, congenital mydriasis, bilateral hemiparesis, swallowing difficulties	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	Yes	Yes
3	9/F/white	PDA, multiple TIAs, hypo-contractile bladder, left hemiatrophy of toes and foot	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	Yes	Yes
4	10/M/white	PDA, congenital mydriasis, unilateral vocal cord paresis, cardiac arrest in newborn period	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	No	Yes
5	10/M/white	PDA, right femoral artery occlusion, dilation aortic arch, pulmonary hypertension	Yes	Yes	Yes	No	Yes	Yes	No	Yes	Yes	Yes
6	4/F/Arabic	PFO, thrombophilia, dystonic left hemiparesis, possible seizures	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	No	Yes
7	3/F/white	Aneurysmal PDA, congenital mydriasis, dilation of ascending aorta	Yes	Yes	Yes	No	Yes	Yes	No	Yes	No	Yes
8	1/F/white	PDA, congenital mydriasis, pulmonary hypertension, deceased due to sepsis	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	No	Yes
9	1/M/Arabic	PDA, congenital mydriasis, bulbar palsy, pulmonary hypertension	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	No	Yes
10	4/F/white	Congenital mydriasis, developmental and speech delays	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	Yes	Yes
11	8/F/Arabic	PDA, congenital mydriasis hands, clumsiness, recurrent TIA	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	No	Yes
12	11/F/white	PDA, aortic dissection, congenital cataracts, cognitive decline	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	No	Yes
13	6/M/white	PDA, congenital mydriasis, left hemiparesis	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	Yes	Yes
14	3/F/white	PDA, congenital mydriasis, recurrent TIAs	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	No	Yes
15[b]	33/F/black	Pseudobulbar palsy, left-sided pyramidal weakness, swallowing difficulties (symptoms started when she was 10 yr of age)	Yes	Yes	Yes (also abnormal posterior temporal gyration in relation to strengthening of PCAs)	No	Yes	Yes	Yes	Yes	No	Yes (few foci)

Note:—PDA indicates patent ductus arteriosus; Pt, patient; PFO, persistent foramen ovale; PCA, posterior cerebellar artery.

[a] Distinctive *ACTA2* cerebrovascular features are dilation of the proximal internal carotid arteries, occlusion of the distal internal carotid arteries, a straight course of arteries of the circle of Willis, and absence of Moyamoya collaterals.

[b] Genetic test was not performed, and clinical context was nonpathognomonic for the *ACTA2* mutation.