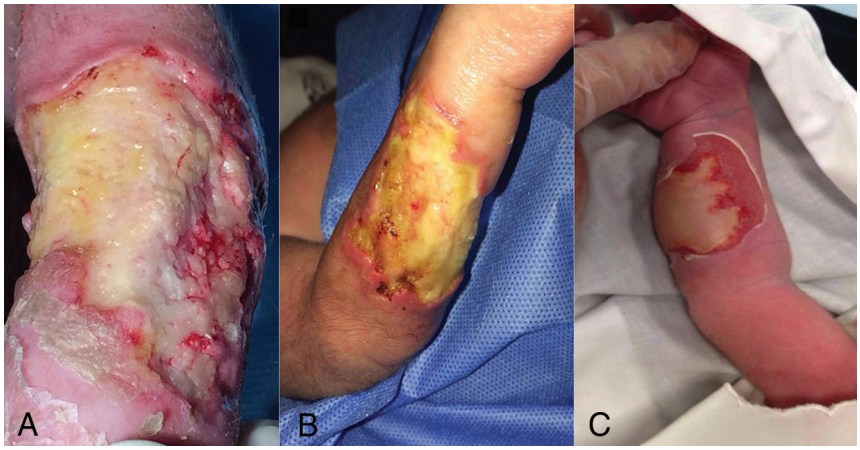
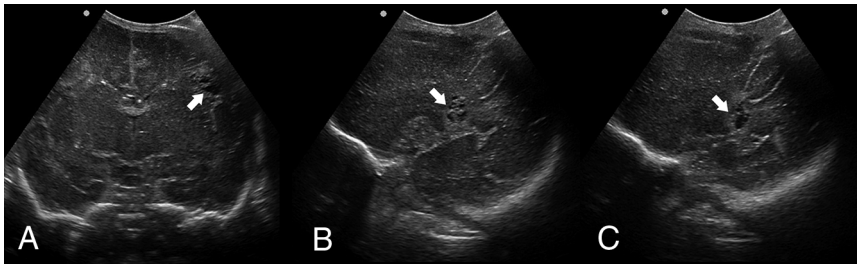
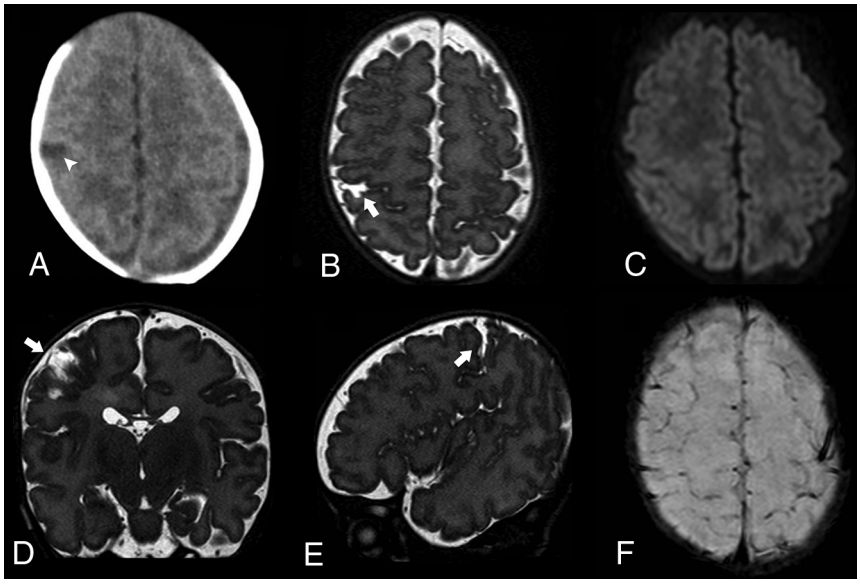


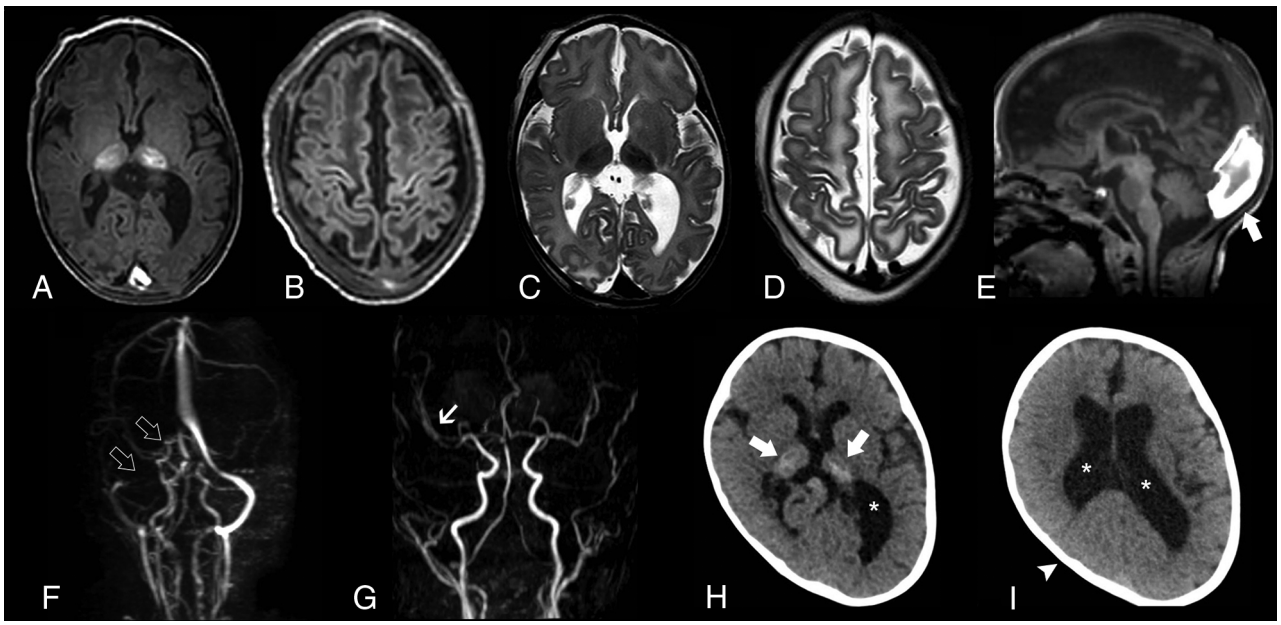
ON-LINE FIG 1. Sequential photographs of the right forearm of patient 1 obtained at birth (*A*) and 2 (*B*) and 4 weeks (*C*) after birth depicting progressive healing of the extensive cutaneous skin ulcer.



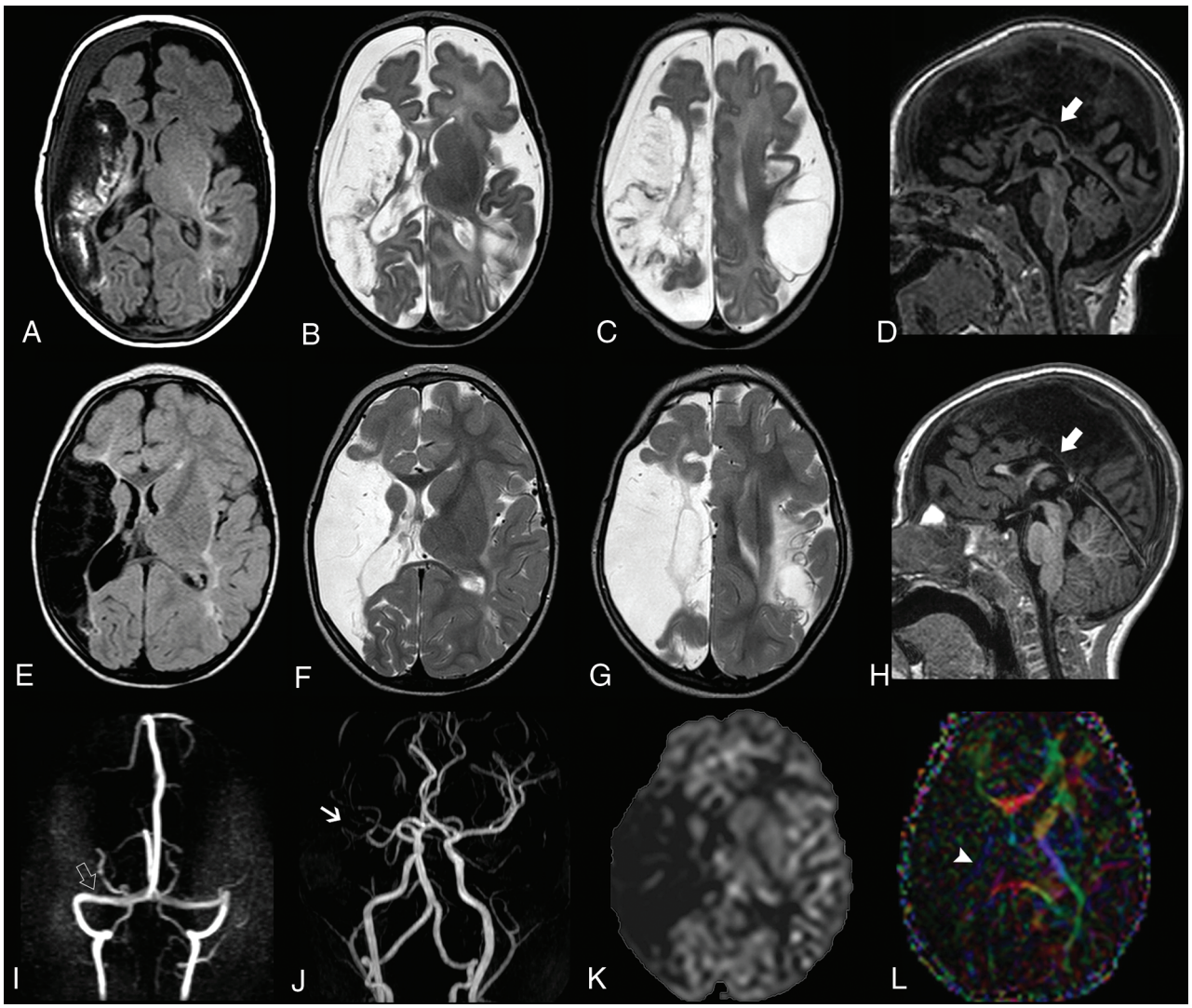
ON-LINE FIG 2. Cerebral sonography of patient 3 on day 1. Coronal (*A*) and parasagittal (*B* and *C*) ultrasonograms depict a focal, irregular area of hypoechogenicity in the subcortical white matter of the right parietal region (*arrow*) compatible with a cavitation in the context of a chronic, prenatal ischemic lesion in the ipsilateral MCA territory.



ON-LINE FIG 3. Neuroimaging studies of patient 4. Unenhanced head CT scan (*A*) performed at 36 gestational weeks of corrected age reveals a focal area of corticosubcortical hypoattenuation in the right parietal region (*arrowhead*). Brain MR imaging obtained at term-equivalent age, including axial (*B*), coronal (*D*), and sagittal (*E*) 3D-driven equilibrium (DRIVE) images confirmed a focal chronic ischemic stroke in the superficial territory of the ipsilateral MCA (*arrow*) causing focal enlargement of the adjacent CSF spaces. Note the absence of associated foci of restricted diffusion on DWI (*C*) or susceptibility artifacts related to hemosiderin deposits on SWI (*F*).



ON-LINE FIG 4. Longitudinal brain MR imaging findings of patient 1. Brain MR imaging performed at 1 month (*A–D*) and at 27 months of age (*E–L*), including axial (*A*) and sagittal (*D*, *H*) T1-weighted images, axial T2-weighted images (*B, C, F,* and *G*), and axial FLAIR (*E*) image reveal progressive atrophic changes over time within the initially affected bilateral cerebral arterial territories and the right thalamus, with ex vacuo dilation of the adjacent CSF spaces. These findings are accompanied by diffuse thinning of the corpus callosum (*arrows*). *I.* MRV, coronal view, performed at last follow-up demonstrates normalization of the caliber and flow-related signal of the torcula and right transverse sinus (*open arrow*). *J.* MRA reveals better representation of the right anterior cerebral artery flow-related signal, with persistent asymmetric MCA branches, less visible on the right (*thin arrow*). *K.* Axial 3D pseudocontinuous arterial spin-labeling map depicts reduced perfusion in the infarcted areas. Axial directionally encoded color fractional anisotropy (FA) map at the level of the basal ganglia (*L*) shows markedly reduced FA of the posterior limb of the right internal capsule (*arrowhead*).



ON-LINE FIG 5. Follow-up neuroimaging of patient 2. Brain MR imaging performed on day 14, including axial (*A* and *B*) and sagittal (*E*) T1-weighted image and axial T2-weighted images (*C* and *D*) show stable appearance of the chronic hypoxic-ischemic encephalopathy signs, as well as reversal of the diffusion abnormalities within the cortical embolic ischemic infarcts in the right MCA territory, likely related to spontaneous recanalization of the distal MCA branches. There is also spontaneous T1 hyperintensity on the torcula, in keeping with dural sinus thrombosis (*arrow*). *F.* MRV, coronal view, confirms absence of flow-related signal in the torcula and right transverse sinus (*empty arrow*). *G.* MRA depicts persistent asymmetry of the MCA branches, less visible on the right (*small arrow*). Unenhanced head CT scan (*H* and *I*) obtained at 9.5 months shows atrophy and spontaneous hyperattenuation of the thalami (*arrows*), as well as mild to moderate ventricular enlargement (*asterisks*) and right positional plagiocephaly (*arrowhead*).

On-line Table 1: Baseline clinical features and neurologic outcome

Patient No.	Sex	IUGR	Prenatal Data	Delivery	GA (wks)	Apgar Scores (1′/5′)	BW, g (SDS)	Length, cm (SDS)	HC, cm (SDS)	Neurologic Presentation	Associated Systemic Findings	UC	FVM Classification	Associated MVM	Age at Last FU	Motor Outcome	Other Neurologic Problems
1	M	No	Threat of preterm delivery	Elective CS	39	8/9	3280 (0.1)	51 (1)	34 (0.1)	Clonic seizures, reduced movements in the R upper limb (day 1)	Skin ulcer	2 turns around the neck at birth	High-grade (first- to second-order stem vessels)	Accelerated villous maturation with hypoxic changes	36 mo	Tetraparesis (L > R)	Microcephaly, severe developmental delay, stereotypies, hyperactivity
2	M	Yes	Twin pregnancy (DD)	Urgent CS	36	6/8	1895 (−2.3)	44 (−1.9)	30.5 (−1.9)	Apneas, clonic seizures (day 1)	Transient myocardial ischemia (day 1)	False knots with torsion	High-grade (amniochorial vessels)	Accelerated villous maturation with hypoxic changes	9 mo (death 10 mo)	Tetraparesis	Microcephaly, severe developmental delay, drug-resistant epilepsy, artificial enteral nutrition
3	M	Yes	Twin pregnancy (DD)	Urgent CS	30	7/8	910 (−2.2)	36 (−1.5)	26 (−1.2)	Incidental finding at US (day 1)	Severe NEC (15 days), RDS (day 1)	Hypocoiled	High-grade (amniochorial and I-II order stem vessels)	Defective implantation with hypoxic changes	NA (death 15 days)	NA	NA
4	M	Yes	Preeclampsia	Urgent CS	30	5/7	930 (−2.2)	35 (−2)	27 (−0.6)	Incidental finding at CT[a] (40 days)	RDS (day 1)	Hypercoiled	High-grade (amniochorial and I-II order stem vessels)	Defective implantation with hypoxic changes	5 yr	Normal	Normal development and cognitive function
5	M	No	PPROM	Urgent CS	36	6/8	2200 (−1.3)	44 (−1.6)	32.5 (−0.1)	Apneas, suspected early sepsis (4 days)	RDS (day 1)	1 turn of bandolier at birth	High-grade (amniochorial and I-IV order stem vessels)	No	6 mo	L hemiparesis	Moderate developmental delay

Note:—BW indicates body weight; CS, cesarean section; DD, dichorionic diamniotic; FU, follow-up; GA, gestational age; HC, head circumference; L, left; M, male; MVM, maternal vascular malperfusion; NA, not applicable; NEC, necrotizing enterocolitis of the neonate; PPROM, preterm premature rupture of membranes; R, right; RDS, respiratory distress syndrome; SDS, standard deviation scores; UC, umbilical cord; US, sonography; IUGR, intrauterine growth restriction.

[a] Performed for suspected choanal stenosis.

On-line Table 2: Neuroimaging characteristics at diagnosis and follow-up

	Patient 1	Patient 2	Patient 3[a]	Patient 4	Patient 5
Cranial sonography	Diffuse WM hyperechogenicity, R > L	Ventriculomegaly, inhomogeneous WM hyperechogenicity	Subcortical WM irregular cavitation, L parietal lobe	Normal	Hyperechoic R periventricular WM
Brain MR imaging (*n*)	6	2	0	1	2
Age at first MR imaging	1 day	1 day	NA	2 mo, 10 days	18 days
Imaging findings at diagnosis					
Infarcts in multiple territories	Yes	Yes	No	No	No
Arterial territories	MCA (R mixed,[b] L superficial), ACA	MCA (superficial, watershed)	MCA (superficial)	MCA (superficial)	MCA (mixed[b])
Laterality	R > L	R > L	L	R	R
Stage	Acute, early, and late subacute	Acute and chronic	Chronic	Chronic	Chronic
Early corticobasal diaschisis	Yes	Yes	NA	No	Yes
Early Wallerian degeneration	Yes (R > L)	No	NA	No	Yes
Associated findings	R subdural fluid collection, dural sinus congestion	Hypoxic-ischemic lesions, dural sinus congestion and thrombosis	NA	No	No
MRA at diagnosis	No signs of arteriopathy, reduced flow signal in R MCA + R ACA	No signs of arteriopathy, reduced flow signal in R MCA	NA	NA	NA
DTI	Asymmetric CST (30 days and 2 yr, 5 mo)	Asymmetric CST (14 days)	NA	NA	Asymmetric CST (6 mo, 26 days)
Age at last MR imaging	2 yr, 5 mo	14 days	NA	NA	6 mo, 26 days
Imaging findings at last FU					
Infarct evolution	Encephalomalacia, gliosis, and atrophy	Reversal of the diffusion abnormalities with minor subcortical residual changes	NA	NA	Encephalomalacia, gliosis and atrophy
New infarcts	No	No	NA	NA	No
Ipsilateral thalamic atrophy and/or signal changes	Yes	Yes	NA	NA	Yes
Ipsilateral CST Wallerian degeneration	Yes	No	NA	NA	Yes
Associated findings	Normalization of dural sinus caliber and venous flow signal	Total recanalization of the dural venous sinus thrombosis	NA	NA	NA
MRA at last FU	Partial improvement	Stability	NA	NA	Reduced flow signal in R MCA

Note:—ACA indicates anterior cerebral artery; CST, corticospinal tract; FU, follow-up; L, left; NA, not available; R, right; MCA, middle cerebral artery.

[a] This preterm neonate died at 15 days of life.

[b] Refers to both superficial and deep territories.