On-line Table: Patient summary[a]

	Neurologic Clinical History	Age	Reported Brain Findings, MR Imaging Unless Otherwise Specified	Reference
New patients				
New patient 1 (Saudi Arabia)	Squint and lower extremity weakness, progressive encephalopathy	4 yr	MR imaging without contrast; lesions in the basal ganglia, brain stem, and cerebellum; enhancing cerebellar lesions	This report
New patient 2 (Saudi Arabia)	Seizure, left arm weakness	8 yr	MR imaging without contrast; multiple focal T2 hyperintense lesions in cerebral and cerebellar cortex	This report
New patient 3 (United States)	Sensorineural hearing loss, progressive CNS findings, esotropia, microcephaly, axial hypotonia, developmental regression, somnolence, seizures, chorea	Prenatal 6 wk 13 mo 15 mo	MR imaging without contrast; subcortical edema, volume loss, T2 hyperintensities in the basal ganglia, internal capsules, ventrolateral thalami, cerebral peduncles, central tegmental tracts and calcifications in globus palladi; progressive edema and necrosis on follow-up	This report
New patient 4 (Turkey)	Rule out brain injury after fall; hypotonia, global developmental delay, ptosis, macrocephaly, incontinence	1 yr 2 yr	MR imaging without contrast;. agenesis of corpus callosum; cerebral cortical atrophy; periventrricular T2 and FLAIR hyperintensity	This report
Previously published patients				
Settas et al[11] 1 (Saudi Arabia)	No neurologic symptoms or findings except for seizure associated with hypoglycemia; family history positive for cousin with hemiparesis and strabismus	5 yr	MR imaging with and without enhancement; foci of T2-FLAIR hyperintensity noted in the basal ganglia, worse in the caudate and anterior putamen, with a decrease in the size of the caudate nuclei; globus pallidus is completely spared bilaterally; intrinsic T1 hyperintensity of the caudate heads; no abnormal contrast enhancement	Settas et al[11]
Bamborschke et al[10] 1 (Germany)	Neonate with microcephaly	3 wk	MR imaging without contrast; generalized cortical atrophy, simplified gyral pattern, hypoplastic temporal lobes, cerebellar hypoplasia; thalamic and hippocampal hyperintensity (T2-FLAIR) extending through the mesencephalon to the pons; no diffusion restriction	Bamborschke et al[10]
Prasad et al[6] 5	Complex partial seizures, developmental delay, failure to thrive, ataxia, sensorineural hearing loss, cranial nerve palsies (III, IV), synkinesia	8 mo 4 yr	MR imaging with and without contrast; contrast enhancement was seen in the globus pallidus, medial thalamic nucleus, and central pons	Prasad et al[6]
Prasad et al[6] 6	Developmental delay, regression, ataxia, hypotonia, sensorineural hearing loss	9 yr	MR imaging with and without contrast; contrast enhancement of cerebellar structures, among other features	Prasad et al[6]

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On-line Table: Continued

	Neurologic Clinical History	Age	Reported Brain Findings, MR Imaging Unless Otherwise Specified	Reference
Lovric et al[5] A544407–306 (Spanish)	None noted	4 yr	Normal CT findings	Lovric et al[5]
Lovric et al[5] A544407–142 (Spanish)	None noted	11 yr	MR imaging with and without contrast; normal MR imaging findings; normal CT findings	Lovric et al[5]
Lovric et al[5] EB1 (Turkey)	Microcephaly	1 mo	MR imaging without contrast; microcephaly, thin corpus callosum, small pons	Lovric et al[5]
Lovric et al[5] B46 (Morocco)	Microcephaly, hypotonia, developmental delay, deafness	3 mo	MR imaging without contrast; microcephaly	Lovric et al[5]
Lovric et al[5] B56 (Morocco)	Microcephaly, hypotonia, developmental delay, deafness	1 mo	MR imaging without contrast; microcephaly	Lovric et al[5]
Lovric et al[5] B1245_21 (European)	Sensorineural hearing loss	3 yr	Normal CT findings	Lovric et al[5]
Lovric et al[5] MC (European)	Peripheral neuropathy (motor and sensory), mononeuritis multiplex, strabismus	18 yr	MR imaging without contrast; normal MR imaging findings	Lovric et al[5]

Note:—EB1 and MC refer to the original designation of individual patients with SPLIS as described in the original reports of cohorts of more than one SPLIS patient.
[a] The numbers in column 1 indicate the reference number of the patient given within the original reference (i.e., how the patient was described in the original report of each series).