

On-line Table 1: Characteristics of pregnancy and MRI data of patients who underwent TOP

Case No./Cyst	Parental History	MRI Findings			Postmortem MRI and Examination
		GA (wk), PVPC (Uni-/Bilaterality, Uni-/Multilocularity, AP Diameter, Height, Typicality, Location)	Additional Findings	Indication for TOP	
21	Prior spontaneous abortion Prior TOP due to increased fetal NT and cerebellar agenesis	35, Bilateral, multilocular, 12 × 6.5 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Dilated temporal horns, porencephalic cyst Additional cyst in the trigon	Porencephalic cyst	MRI: ventricular border could not be identified due to ventricular collapse Postmortem: irregular cystic lesions parallel to the ventricles; PVPC in the right temporal horn; missing ependymal, replaced with gliosis in the left occipital horn
22	—	35, Bilateral, multilocular, 10 × 6 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Asymmetric and moderately dilated lateral ventricles T2 hyperintense signal in the white matter and irregular temporal horns	Suspected fetal ventriculitis	MRI: bilateral PVPC adjacent to the frontal horns, smaller than demonstrated in the uterus Postmortem: bilateral PVPC associated with edema, gliosis, and increased microglia No postmortem due to fetal reduction
23	2 Prior spontaneous abortions Maternal and fetal CMV infection during pregnancy Twin pregnancy, fetal reduction	29, Bilateral, multilocular, 3.5 × 4 mm, atypical, adjacent to the frontal horns, anterior to the caudothalamic notch	—	Fetal CMV infection	
24	Subependymal pseudocysts Daughter with developmental delay, congenital cataract, nystagmus, hypotonia, and seizures	31, Bilateral, multilocular, 10.3 × 6.5 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Mildly dilated lateral ventricles; small porencephalic lesion in the right frontal horn, attached to the lateral ventricle	Porencephalic cyst	MRI: dilated right lateral ventricle with adjacent porencephalic cyst and hypointense signal in T2 series compatible with hemorrhage Postmortem: white matter necrosis in the right fronto-parietal lobe with porencephalic cyst TOP in another hospital
25	Mother, Kallmann syndrome Maternal and fetal CMV infection during pregnancy	33, Bilateral, multilocular, 10.5 × 6 mm, typical, adjacent to the frontal and occipital horns, anterior and posterior to the caudothalamic notch	Asymmetric lateral ventricles T2 hyperintense signal in the white matter in both hemispheres	Fetal CMV infection	
26	Polyhydramnios	29, Bilateral, multilocular, 6.6 × 4.8 mm, typical, adjacent to the frontal horns, anterior and posterior to the caudothalamic notch	Evidence for bilateral germinal matrix hemorrhage Mildly dilated lateral ventricles	PVPC with additional dilated lateral ventricles and germinal matrix hemorrhage	TOP in another hospital

Note:—NT indicates nuchal translucency; AP, anteroposterior; GA, gestational age.

On-line Table 2: Characteristics of pregnancy, MRI data, and neurodevelopmental outcome of children with isolated PVPC (group A)

Case No./Cyst	Parental History	MRI Findings, GA at Time of MRI (wk), PVPC (Uni-/Bilaterality, Uni-/Multilocularity, AP Diameter, Height, Typicality, Location)	Neurodevelopmental Outcome (Age)
Connatal cysts			
1	Prior child with choroid plexus cysts	34, Bilateral, multilocular, 8.5 × 6 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Adequate adaptive level according to Vineland II (3 yr 8 mo)
2	Antiplatelet therapy during pregnancy; heterozygote to <i>MTHFR</i> and <i>FVL</i> mutations Prior TOP due to PVPC finding Prior pregnancy with PET	33, Bilateral, multilocular, 7.5 × 5.8 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Clinical follow-up: normal development with mild hypertonía (10 mo); adequate adaptive level according to Vineland II (3 yr 6 mo)
3		33, Bilateral, multilocular, 14.5 × 7 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Clinical follow-up: normal findings (9 mo) Adequate adaptive level according to Vineland II (2 yr 4 mo)
4	APLA syndrome, heterozygote to factor 2 mutation Prior child with cerebral palsy Prior TOP due to multiple fetal malformations Maternal CMV infection during pregnancy (negative serology in amniocentesis) Twin pregnancy, TTTS, fetal reduction	33, Bilateral, multilocular, 6 × 4.5 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Urine test negative for CMV after birth Clinical follow-up: mild hypotonía (3 mo), normal examination findings without hypotonía (6 mo) Adequate adaptive level according to Vineland II (1 yr 3 mo)
5		32, Bilateral, multilocular, 7 × 5 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Clinical follow-up: torticollis and hypotonía (3 mo), normal findings (6 mo) Adequate adaptive level according to Vineland II (1 yr 1 mo)
6	Homozygote to <i>MTHFR</i> mutation, heterozygote to <i>FVL</i> mutation, low C protein levels Antiplatelet and antipsychotic treatment during pregnancy Prior pregnancy with PET	33, Bilateral, multilocular, 8.7 × 5.5 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Adequate adaptive level according to Vineland II (1 yr 1 mo)
Subependymal pseudocysts			
7	Maternal toxoplasma infection during pregnancy (no amniocentesis)	34, unilateral (right), unilocular, 11.5 × 5.7 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Adequate adaptive level according to Vineland II (1 yr 6 mo)
8	Mother, hypothyroidism Two prior spontaneous abortions Prior child with DENT disease and <i>CLCN5</i> mutation Prior child who died at the 14 mo of age from pneumonia Polyhydramnios	31, Bilateral, multilocular, 8.5 × 6.5 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Negative saliva test for CMV after birth Clinical follow-up: healthy (3, 9, 12 mo) Adequate adaptive level according to Vineland II (1 yr)

Note:—TTTS indicates twin-to-twin transfusion syndrome; PET, pre-eclamptic toxemia; APLA, antiphospholipid antibody AP, anteroposterior; GA, gestational age.

On-line Table 3: Characteristics of pregnancy, MRI data, and neurodevelopmental outcome of children with nonisolated PVPC and normal outcome

Case No./Cyst	Parental History	MRI Findings			Neurodevelopmental Outcome (Age)
		GA (wk), PVPC (Uni-/Bilaterality, Uni-/Multilocularity, AP Diameter, Height, Typicality, Location)	Additional Findings		
9	—	35, Bilateral, unilocular, 8.5 × 3.3 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Asymmetric and moderately dilated lateral ventricles	Adequate adaptive level according to Vineland II (2 yr 11 mo)	
10	Father, CSP1 deficiency Prior spontaneous abortion	35, Bilateral, multilocular, 11 × 5.2 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Asymmetric lateral ventricles	Clinical follow-up at 3, 6, 9 mo, 1 yr, 1 yr 3 mo: at 3 mo, mild developmental delay, hypotonia, and torticollis; at 1 yr 3 mo, normal development	
11	Prior spontaneous abortion Maternal family, spina bifida, familial dysautonomia Paternal family, mental retardation SSRI treatment during pregnancy	31, Bilateral, multilocular, 6 × 4.6 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Asymmetric and mildly dilated lateral ventricle	Moderately high adaptive level according to Vineland II (2 yr 6 mo) Moderately high adaptive level according to Vineland II (2 yr 4 mo)	
12	SSRI treatment during pregnancy IUGR	37, Bilateral, multilocular, 6.2 × 3.7 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	—	Moderately high adaptive level according to Vineland II (1 yr 9 mo)	
13	PVPC in prior pregnancy	32, Bilateral, multilocular, 8 × 5.5 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Asymmetric lateral ventricles	Clinical follow-up: healthy (9 mo) Adequate adaptive level according to Vineland II (1 yr 6 mo)	
14	IUGR	37, Bilateral, multilocular, 10.5 × 5 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Asymmetric lateral ventricles; suspected T2 hyperintense signal in the white matter	Clinical follow-up: healthy (9 mo) Adequate adaptive level according to Vineland II (1 yr 5 mo)	
15	Subependymal pseudocysts Antiplatelet treatment due to a TIA during pregnancy Maternal CMV infection during pregnancy (negative serology in amniocentesis)	34, Unilateral (left), multilocular, 8.5 × 5.5 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	T2 hyperintense signal in the white matter	Clinical follow-up: healthy (3 mo) Adequate adaptive level according to Vineland II (2 yr 4 mo)	
16	Father, myotonic dystrophy Prior child with white matter disease, died at 4 yr IUGR	36, Bilateral, multilocular, 13 × 5.5 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	T2 hyperintense signal in the white matter lobes	Adequate adaptive level according to Vineland II (3 yr 8 mo)	
17	Prior spontaneous abortion	38, Bilateral, multilocular, 8.5 × 6 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Asymmetric and moderately dilated lateral ventricles, dolichocephalic skull	Moderately high adaptive level according to Vineland II (2 yr 6 mo)	
18	Antiepileptic treatment during pregnancy Maternal family, mental retardation Paternal family, epilepsy	38, Bilateral, multilocular, 7.2 × 5.2 mm, typical, adjacent to the frontal horns, anterior and posterior to the caudothalamic notch	Dilated temporal horns; T2 hyperintense signal in the white matter; asymmetric and mildly dilated lateral ventricles; rotation of the vermis without a correct closure of the 4th ventricle	Saliva test negative for CMV after birth Clinical follow-up: healthy (2, 5 mo) Adequate adaptive level according to Vineland II (1 yr 2 mo)	

Note:—SSRI indicates selective serotonin reuptake inhibitor; AP, anteroposterior; GA, gestational age.

On-line Table 4: Characteristics of pregnancy, MRI data, and neurodevelopmental outcome of children with nonisolated PVPC and abnormal outcome

Case No./Cyst	Parental History	MRI Findings			Fetal Abnormalities	Neurodevelopmental Outcome (Age)
		GA (wk), PVPC (Uni-/Bilaterality, AP Diameter, Height, Typicality, Location)	Additional Findings			
Subependymal pseudocysts 19	—	29, Bilateral, multilocular, 6.5 × 4.5 mm, typical, adjacent to the frontal horns, anterior and posterior to the caudothalamic notch	Asymmetric and moderately dilated lateral ventricles; T2 hyperintense signal in the white matter and sulcation abnormality	1P36 microdeletion	Developmental delay, vision problems and seizures Low adaptive level according to Vineland II (2 yr 2 mo)	
20	Prior spontaneous abortion	32, Bilateral, multilocular, 7.6 × 5.6 mm, typical, adjacent to the frontal horns, anterior to the caudothalamic notch	Cerebellum measurement (TCD) <3rd percentile without abnormality in its structure; abnormal structure of the 4th ventricle	IUGR Abnormal echo test findings, VSD and suspected AV canal Translocation in chromosomes 11 and 22	Low adaptive level according to Vineland II (1 yr 9 mo) Developmental delay; Emanuel syndrome (OMIM No. 609029)	

Note:—YSD indicates ventricular septal defect; AV, atrioventricular; TCD, transverse cerebellar diameter; OMIM, Online Mendelian Inheritance in Man; AP, anteroposterior; GA, gestational age.

On-line Table 5: Vineland II adaptive behavior composite standard scores

Group/Case No.	Age	Adaptive Behavior Composite Standard Score	Adaptive Level	Percentile Rank
A				
1	3 yr 8 mo	102	Adequate	55
2	3 yr 6 mo	114	Adequate	82
3	2 yr 4 mo	102	Adequate	55
4	1 yr 3 mo	100	Adequate	50
5	1 yr 1 mo	106	Adequate	66
6	1 yr 1 mo	105	Adequate	63
7	1 yr 6 mo	98	Adequate	45
8	1 yr	95	Adequate	37
B				
9	2 yr 11 mo	100	Adequate	50
10	2 yr 6 mo	121	Moderately high	92
11	2 yr 4 mo	122	Moderately high	93
12	1 yr 9 mo	115	Moderately high	84
13	1 yr 6 mo	105	Adequate	63
14	1 yr 5 mo	108	Adequate	70
15	2 yr 4 mo	98	Adequate	45
16	3 yr 8 mo	103	Adequate	58
17	2 yr 6 mo	116	Moderately high	86
18	1 yr 2 mo	98	Adequate	45
19	2 yr 2 mo	58	Low	<1
20	1 yr 9 mo	63	Low	1