

WEST SYNDROME AND ZIKA VIRUS INFECTION IN PRENATALLY EXPOSED CHILDREN: UNEXPECTED ASSOCIATED PARTNERS?

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Background:

- Zika virus (ZIKV) has been associated to microcephaly and other neuro-developmental abnormalities.
- West syndrome (WS) is a severe epilepsy composed of the triad of infantile spasms, hypsarrhythmic electroencephalogram (EEG) pattern, and mental retardation.
- ZIKV laboratory diagnosis in pregnant women is difficult in low-middle resource countries.
- Overall, 143 infants born to suspected ZIKV-infected mothers are followed-up in a tertiary paediatric hospital in Guayaquil (Ecuador).
- We report the diagnostic and treatment challenges of first four cases of WS associated to ZCS.

Cases Presentation Summary:

- All the reported cases had Zika Congenital Syndrome (ZCS) signs/symptoms, epileptic seizures and hypsarrhythmic EEG pattern compatible with WS (table 1).
- All the mothers, except case 4, had no laboratory evidence for ZIKV infection during their pregnancy.
- CSF and/or urine tested negative for CMV, *Toxoplasma gondii*, and ZIKV in all the children.
- In all the cases, seizures didn't response to valproate, and vigabatrin as compassionate medication controlled them.
- This drug had to be obtained from foreign country.
- Case 4 presented other congenital malformations associated to ZCS such as brachyoligodactyly and dysphagia (figures 1, 2, 3, and 4).

Case	Sex (M/F)	Mother's symptoms during pregnancy (yes/no)	GA at birth (weeks)	HC at birth (cm/z-score)	Microcephaly (yes/no)	Age at first seizures (months)	Age at diagnosis of WS (months)	Brain CT scan
1	M	yes	39	31(-3.4)	Yes	4	12	ventriculomegaly Calcifications in basal ganglia
2	M	no	40	32(-2.5)	Yes	3	11	ventriculomegaly, thin cortex, and calcifications in basal ganglia
3	F	no	40	27.5(-6.4)	Yes	6	9	Subcortical calcifications and pachygyria
4	F	yes	38	32(-2.0)	yes	6	7	Ventriculomegaly and pachygyria

Table 1. reported cases with their principal characteristics regarding ZIKV infection

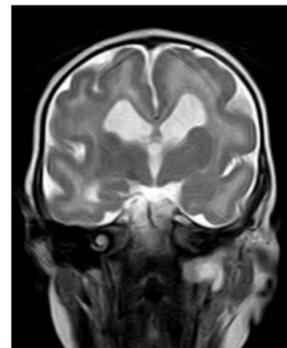


Figure 1. Brain MRI with ventriculomegaly and pachygyria

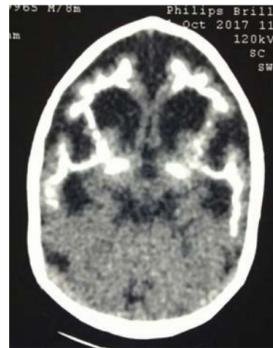


Figure 2. Brain CT scan with subcortical calcifications

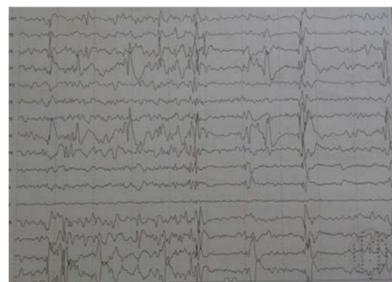


Figure 3. Modified hypsarrhythmic EEG pattern



Figure 4. Phenotypic aspect of Congenital Zika Syndrome

Learning Points/Discussion

- Lack of diagnosis in ZIKV-infected mothers during pregnancy is a great challenge for low-middle resource countries.
- We here report the first four cases of WS associated to ZCS diagnosed in our country Ecuador.
- The delay for its diagnosis could result in severe adverse neurological outcomes for these babies.
- Moreover, the absence of vigabatrin in the country worsened the critical clinical situation of these children.