TIC-TACK-TOE,
THREE IN A ROW!
WHY THIS WOE?!!!

Prof. R.C.M (M 7)UNIT.
ICH and HC

Dr.G.THANNOLI GOWTHAMI
Post Graduate in M.D Paediatrics

Complaints

- 3 year old girl child
- 2nd born of 3*consanguinity

FTND

No perinatal insult

Developmentally – Normal

Unsteadiness of gait - 10 days

Present illness

Presented with acute onset of unsteadiness of gait noted while walking and standing- 10 days

Clumsiness and jerky movements of UL, Trunk and LL

Abnormal eye movements

- Slurring of speech+
- progressive in nature
- H/o Irritability + No H/o vomiting & seizures

History

No history suggestive of ear discharge ,toxin exposure,trauma, exanthematous fever,rash, No h/o any GI or URI
No H/o recent vaccination

Examination

On admission:

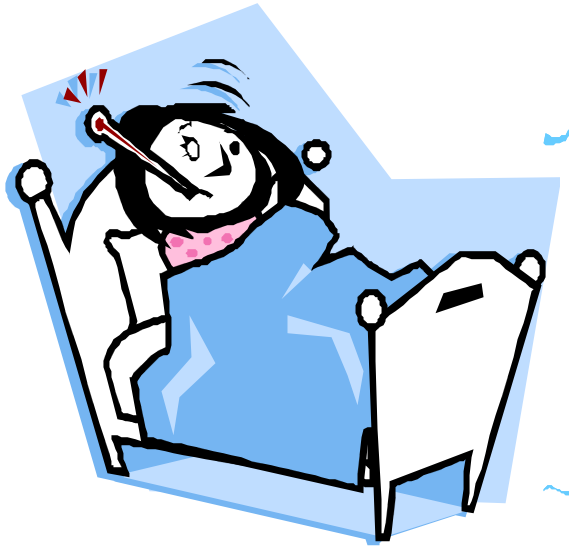
General appearance: irritable,

Afebrile,anicteric and not anemic.

Vital signs:

RR = 28/min, Pulse = 100 beats/min.

Weight = 12 Kgs, Height = 88 cms.



CVS: Normal

RS: Vesicular breath sounds heard.

ABD:Soft,No organomegaly

Examination

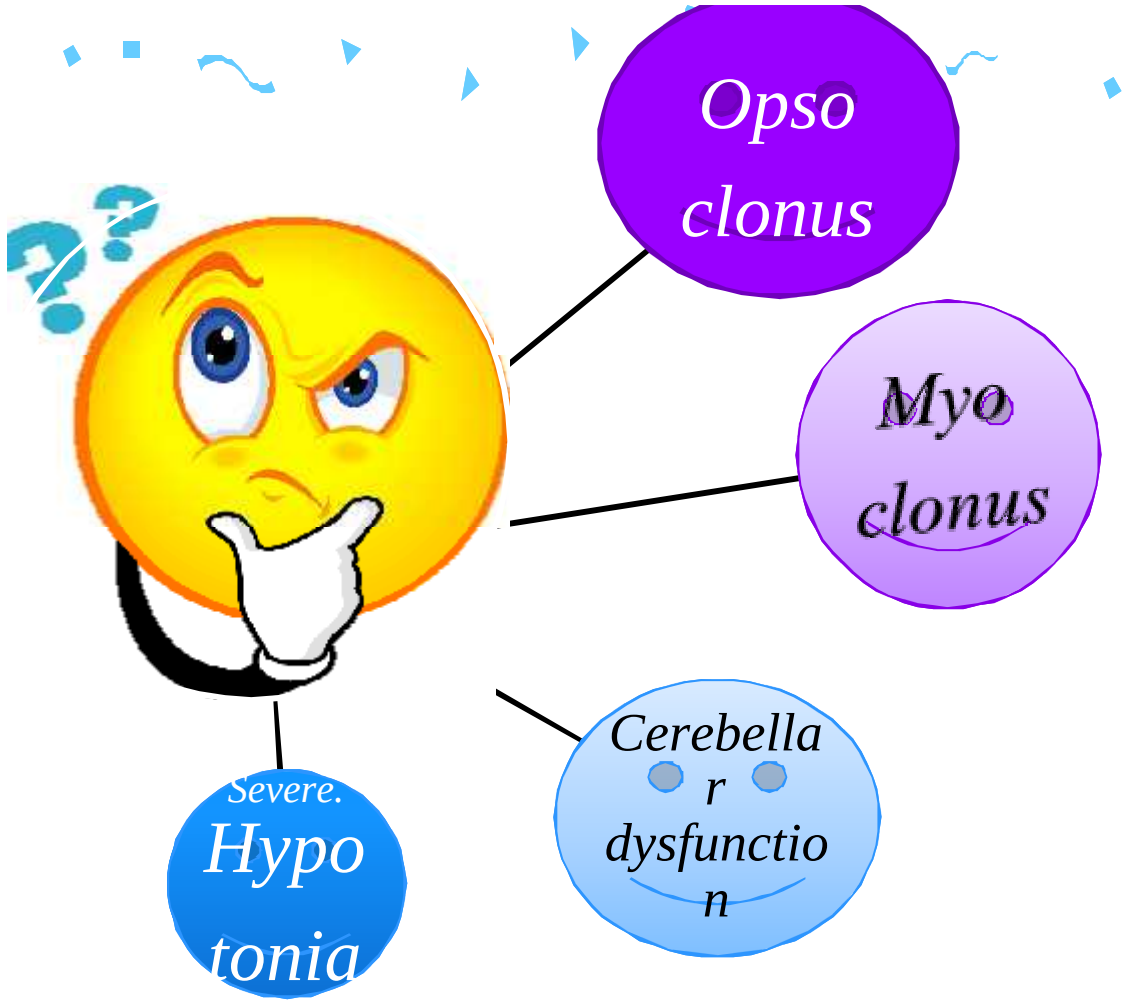
Child irritable, PERL, EOM full, Opsoclonus, Head lag,

Titubation,

Truncal ataxia

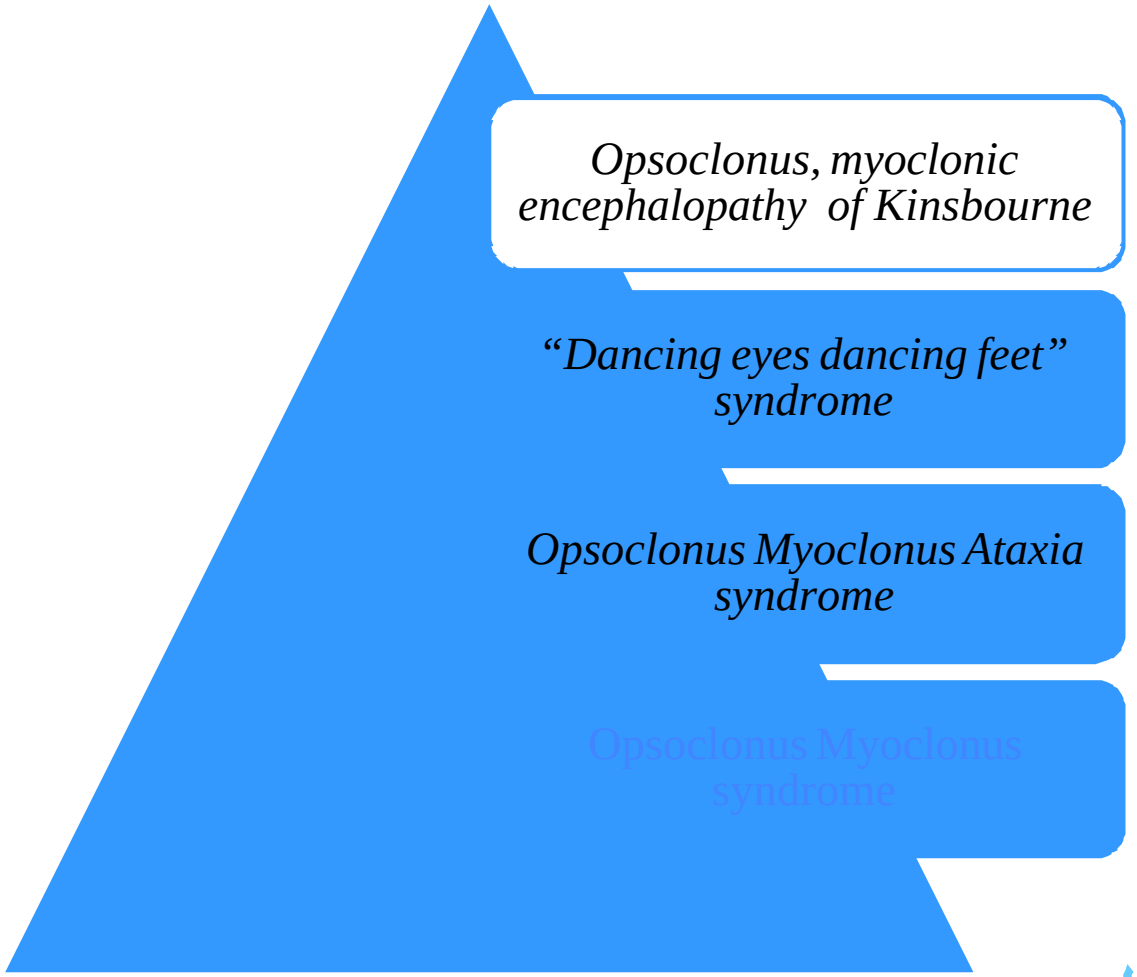
Myoclonic jerks involving limbs and trunk

Hypotonia

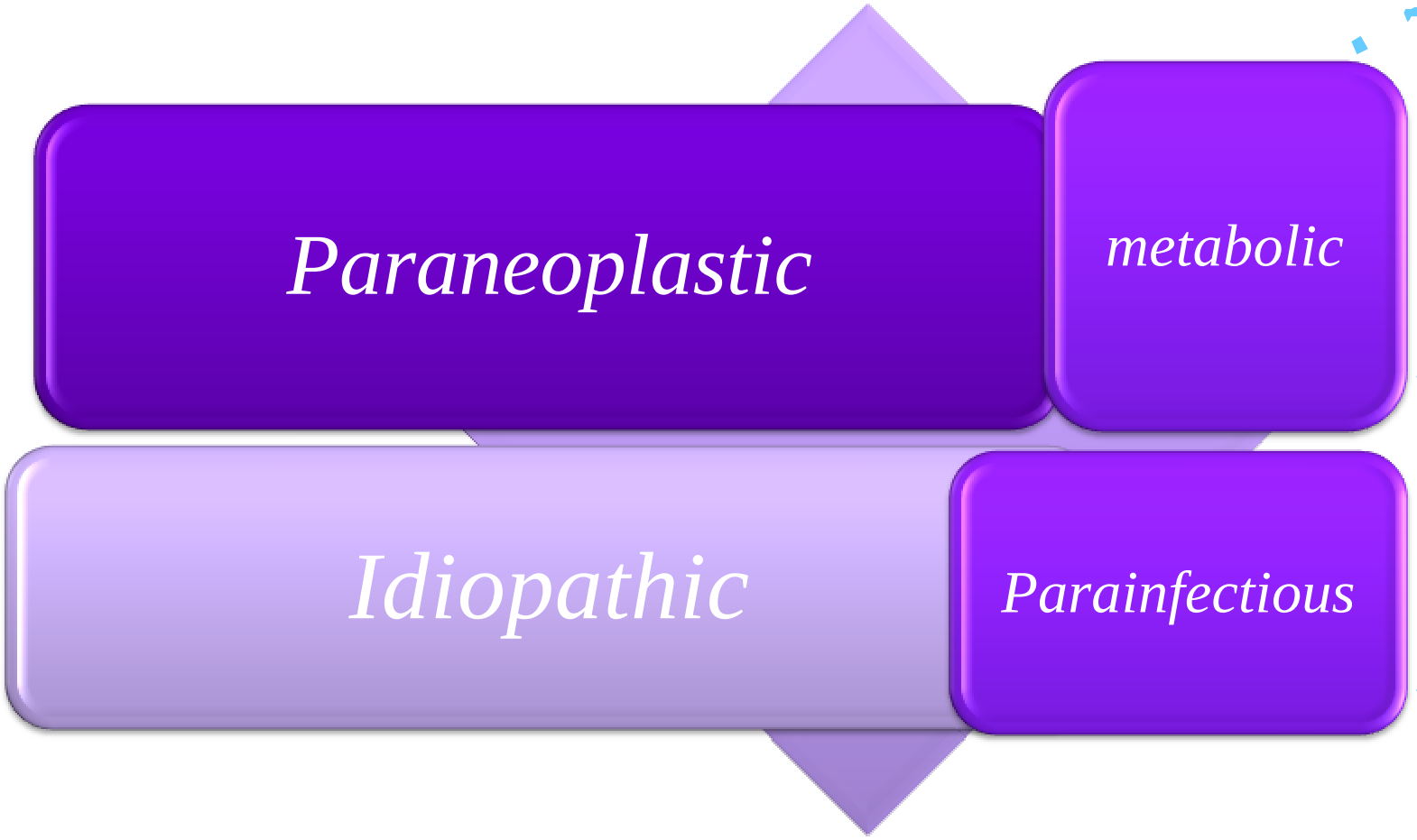


Opsoclonus Myoclonus syndrome

Opsoclonus Myoclonus syndrome

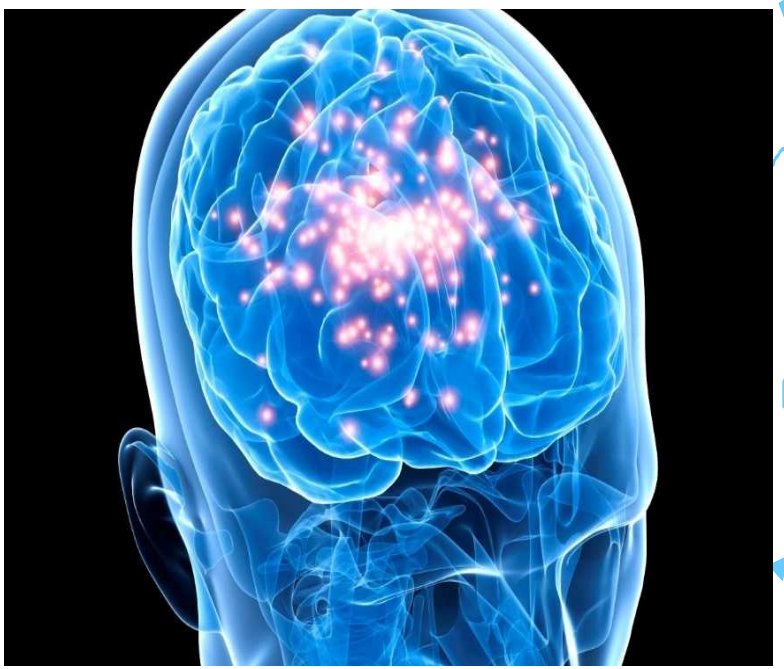


Aetiology



Opsoclonus Myoclonus syndrome

The pathophysiology of OMS has been speculated to involve IgG and IgM autoantibodies directed against neural antigens in cerebellar Purkinje cells, cerebral cortical neurons and axons.



Outcome and Prognostic Features in Opsoclonus-Myoclonus Syndrome From Infancy to Adult Life

Andreas Brunklaus, Keith Pohl, Sameer M. Zuberi and Carlos de Sousa

Pediatrics 2011;128;e388; originally published online July 25, 2011;
DOI: 10.1542/peds.2010-3114

Chronic relapsing

Motor abnormality/ataxia

Opsoclonus (intermittent)

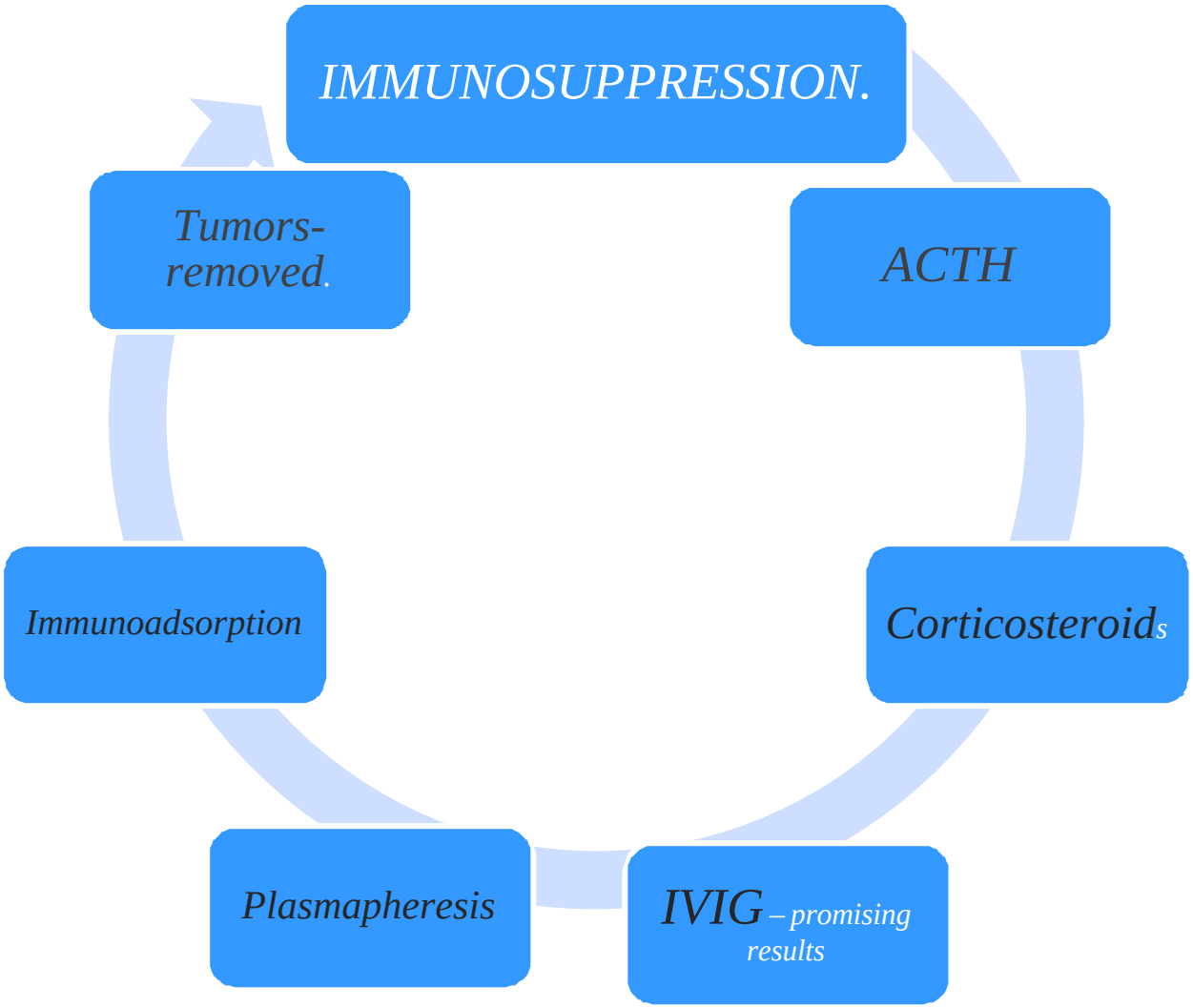
Speech abnormality

Learning disability

Behavior problems

Ongoing medication

OMS -Treatment



Investigations

CBC	*Investigations*	
TC	7100 cells	
DC	P40	L60
Hb	12.7G%	PCV: 36 %
Platelets	2 lakhs	
LFT		
Bilirubin <1mg	SGOT 28 u	SGPT 19 u
RFT		
Urea 16mg	Creatinine 0.8 mg	
Sr.Electolytes	Na 136meq K 4meq	Hco3 19meq
Blood Sugar 95 mg®	Ca 9.2 mg	
Urine Routine – N	Urine C/S	No growth
CXR Normal	USG Abd and neck	Normal

Further investigations

Test	Value	Test	
Sr.Lactate	*36*	Urine VMA	5.56 N
Sr.Ammonia	*58 mic/dl*	Urine VMA/Creat	12.6 N
Urine MS	*Negative*	CSF Glucose	66mg%
Sr.Uric acid	*4.1mg*	Protein	16mg%
Vit B12	*711 mg*	Cell count	3 cells
		Viral studies	Negative
TMS	*Normal*	Latex Agg test	Negative
TFT	*Normal*	Gram stain	Negative
EEG	*Normal record*	HIV, Hep B,HCV	Negative
MRI Brain	*Normal*	CT chest,CT abd	Normal

Treatment

Iv methyl prednisolone,,
ACTH,
IVIG,
Oral prednisolone,
Sodium valproate and Clonazepam

WHAT'S KNOWN ON THIS SUBJECT

Opsoclonus-myoclonus syndrome (OMS) in children

- *chronic-relapsing*
- *debilitating illness of early childhood*
- *atypical presentation.*

Children with severe initial symptoms and those who are very young at disease onset are at particular risk of developing long-term neurologic sequelae.

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Take Home Message

It is important for those affected to be identified early, because they might benefit from new advances in immunomodulating and immunosupressive therapy. Follow up is mandatory.

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OMS

ICH data (10 years) 2004 -till date

Total - 7 cases

3 - paraneoplastic

1- neurodegenerative

1- infective,

2 -idiopathic

Teachers !
They inspire you , they entertain you, you end up learning a ton even when you do not know it !

Thank you

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Prof. Leema Pauline

Asst Prof.

Dr.Viveka Saravanan

Dr.Ravi

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Video

Diagnosis

Opsoclonus Myoclonus syndrome

The overall incidence of OMS in children with neuroblastoma is approximately 3%.

50% patients -idiopathic cause

OMS -neuroblastoma, ganglioneuromas ,hepatoblastoma.