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AN ANNOTATED COMPILATION OF CHILDHOOD PERVASIVE DISINTEGRATIVE SPECTRUM DISORDERS(PDD) CHECK LISTS WITH EMPHASIS ON DIFFERENTIAL ALGORITHMIC,DIAGNOSTIC THERAPEUTIC-INTERVENTIONAL APPROACH

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Background-Historical-notes-purpose: PDDs are-gradable-constellations of neurolobiological-disorders with abbreviated-life -expectancies, including CDD,its anecdotal-extreme-rarity made it initially unscientifically-acknowledged-Retts-syndrome(RS)-affects only females.Previously most girls diagnosed asCDD,retrospectively had RS.In addition to Autism-spectrum-disorders(ASD),-Aspergers syndrome(AS).An unequivocal initial diagnosis of a specific PDD was unusual,because of their striking overlapping features.Although features were spectral-overlapping,they were aetiologically-prognostically distinct.Accurate-diagnosis is crucial because interventions-differ. The composite-roles of prematurities-birth- traumas-genetic factors-amyloid-interleukins-1-beta-endorphins-environmental- toxigenic- autoimmunity-related disruption of neuronal-transmissions-neuro-pathological associations with TSC, -intrauterineTORCHES-viral-exanthematous-related-SSPE-associated-symptomatic-seizures which suggests the-diagnosis of otherwise-similar-but- very -therapeutically-responsive-Landau-Kleffner-syndrome(LKS) especially in boys-the influence of other-infective-processes suggests the benefits of an apposite index of suspicion for a timely diagnosis/-specific-interventions.Ongoing research although undermined by the relative-rarity of these disorders-suggests that overall,PDDs-were underdiagnosed-misdiagnosed.However the putative-roles of infective-antecedents suggests that impacts could be epidemio-geographically distinct.

A delineation of categorical-figures from a defined-setting-could be scientifically -illuminating with implications for research-directions.

Methods-case-definitions-interventions: Descriptors- pre-morbid-post morbid- historical notes were retrieved from teachers-care givers-parents of pervasive-developmental-educational-social-emotional- physical aspects of cases-relevant to the theme.Putatively-universally-acceptable-symptomatic-syndrome-driven-classifiers with batteries of developmental-tests were applied for

Case-definitions.Interventions were structured-behavioural-educational-speech-language therapies-social skills-development-sensory-integration- occupational therapies-hypnotic-sedatives-antiepileptics-tranqulizers.Rewards to reinforce desirable behaviours-discouraging-untoward-behaviours-Cares in respite-homes were proffered.

Results: Of the(n=13)cases-(n=8) were males. Mean-age at diagnosis =89.08months.

On the basis of epidemiological-historical-demographics-temporality-chronology-interventional-responsiveness of symptomatologies,the diagnosis of compatible-early infantile-autism-spectrum-disorders was suggested in(n=1),compatible high functioning autism.(n=1)CDD(n=1),Retts-syndrome(n=1). PPD-NOS (n=9)-no compatible- AS.Unprovoked-Remote-symptomatic-seizures were frequent(n=11) Other associations were primary enuresis-social-deviance-defective-self-regulation-deranged-sleep-wake cycles-acute-psychosis-inordinate-temper-tantrums-non-specific-abdominal-pains-hallucinations-recurrent- febrile-responses-irritable-bowel-syndromes-multiphasic-hallucinations-poor co-operation-attention span-concentrations/-syncope.

Conclusions: In PPD, because outcomes were improved with earlier interventions,an opportunistic screening for developmental-defects should be undertaken in every well baby-child visits.Although symptomatically similar,- patterns of onset-course-outcomes were categorically distinct.A consistent-positive approach results in improved outcomes. These figures could direct-diagnostic-therapeutic-approaches/-relevant interventions.

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EFFECT OF NEONATAL HYPOXIA/ISCHEMIA ON GABA$_A$ RECEPTOR PROTEIN EXPRESSION

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Background and aims: Current therapeutic options for neonatal hypoxia/ischemia (HI) are

limited to hypothermia and treatment of seizures. The principal function of the GABA system in mature brain is inhibition, however in neonatal brain GABA provides much of the excitatory drive. Thus whilst anticonvulsants augment GABA's inhibitory actions in mature brain, administration of GABAergic drugs to neonates may potentially exacerbate seizures and worsen HI brain injury. Furthermore changes in $GABA_A$ receptor expression will influence receptor pharmacology. We aimed to assess changes in protein expression of the $GABA_A$ receptor in the neonatal HI piglet.

Methods: Newborn piglets were subjected to a 30 min HI insult and euthanased at 72 h. Brain tissue was collected from several brain regions and $GABA_A$ receptor α_3 protein expression levels analysed by western blot.

Results: Expression of α_3 was significantly elevated in frontal cortex of HI animals without seizure; animals with seizure trended toward lower a_3 expression. In occipital cortex, α_3 expression was found to be diminished in all HI animals however presence of seizures was associated with significantly lower α_3 expression.

Conclusions: $GABA_A$ receptor α_3 expression was significantly altered following neonatal HI; presence of seizures further changed this expression. Efficacy of anticonvulsants in neonatal brain may not only depend on regional and temporal maturation of GABAergic inhibitory function but also on $GABA_A$ receptor subunit expression following both HI and seizures. There is a critical need to develop effective treatment strategies specific to the neonatal HI brain.

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ROUTINE MAGNETIC RESONANCE IMAGING OF THE BRAIN AT TERM CORRECTED AGE IN ALL OUR VERY LOW BIRTH WEIGHT INFANTS

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Background and aims: To compare findings of cranial ultrasound (US) and magnetic resonance imaging (MRI) of the neonatal brain.

Methods: A retrospective population based cohort study of premature infants with a very low birth weight (VLBW) of less then 1500 grams who were admitted to our neonatal unit on day 1 (period: 2007 to 2009).

All infants underwent cranial US on day 1, 3, 7, 14, 28, 42 monthly thereafter and at term corrected age. Infants who were eligible for this study obtained a MRI at term (37 to 42 weeks gestational age) corrected age.

Retrospectively, we compared the cranial US findings as a predictor of a wide spectrum of pathology on MRI.

Results: Paired MRI and US studies were performed in (n=140) VLBW infants who were born at a median gestational age of 28 (range: 22+1 to 34+5) weeks and a median birth weight of 1020 (range: 335 to 1495) grams.

US predicted some MRI findings accurately: germinal layer haemorrhage (GLH), cystic lesions, intraventricular haemorrhage (IVH) and severe white matter (WM) echogenicity on US for the presence of WM haemorrhagic parenchymal infarction on MRI.

Other MRI changes were less well-predicted: delay in maturation and myelination, reduced cortical folding, congenital malformations, punctate lesions and mild or no WM echogenicity on US for the presence of normal (n=112) WM signal intensity on MRI.

Conclusions: MRI of the neonatal brain might shed light on the origin of brain lesions causing long-term neurodevelopmental sequelae and might change our perinatal and neonatal management.

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NEUROMUSCULAR ACTIVATION DURING REVERSE AND NON-REVERSE CHEWING CYCLES IN CHILDREN WITH UNILATERAL POSTERIOR CROSSBITE

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Introduction: Posterior unilateral crossbite is an asymmetric malocclusion developing early, during