Neurocognitive-Affective Dysfunction in Dandy Walker Malformation involving the Cerebellum: A Case Study

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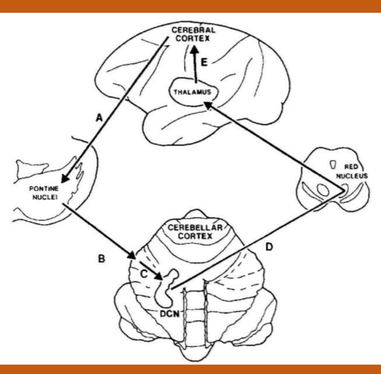
Background

- **Damage to the cerebellum has been associated with cognitive and emotional deficits[1]**
  - The Dandy Walker Malformation (DWM) is a congenital defect characterized by cerebellar dysgenesis, 4th ventricular dilation, and enlarged posterior fossa[2]
  - The role of the cerebellum in DWM, may have additional implications on non-motor functions
  - **Cerebellar Cognitive Affective Syndrome** (CCAS)[3]: Perseveration, inattention, visuospatial defects & affective and personality changes

Anatomical circuitry linking cerebellum and cerebral cortex[4]



Patient

- **48-year-old, right-handed Caucasian man, diagnosed at birth with DWM**
  - Underwent 4th ventriculosomy surgery at 3 months
- **Concerns:** **No complaints by patient;** Father concerned about son's current functioning and ability to care for self n future
  - Unstable employment and housing; financial dependence
  - Patient less independent and self sufficient than peers
- **Developmental milestones:** WNL except for walking (age 2)
- **Education**: Bachelor's degree ; Denied learning/ ADHD
- **Psychosocial:** Never married; a few friends
- **Medical-Psychiatric History**: Unremarkable other than DWM
- **Medications:** None
- **MRI Findings (2014):** Enlarged 4th ventricle, incomplete vermis
- **Behavioral Observations:** Cooperative and attention, but rigid and somewhat restricted during interactions

Test Findings

Intellect	Raw	SS	%ile
WAIS-IV Full Scale IQ	121	114	82
Verbal Comp (VCI)	45	130	98
Percep Reason (PRI)	28	96	39
Working Mem (WMI)	30	128	97
Processing Speed (PSI)	12	94	34

Front-Motor	Raw	T-Score	%ile
Grooved Pegboard-Right	92	33	5
Left	101	33	5
Luria Contrasting	0 errors		WNL
Luria Go-No-Go	0 errors		WNL

Facial Affect	Raw	T Score	%ile
Florida Affect Battery			
Facial Identity	18	-	95
Facial Affect Discrim	19	-	90
Naming Affect	19	-	95
Matching Affect	17	-	85

Mood	Raw	T-Score	%ile
Beck Depression Inventory	0		
State-Trait Anxiety- State	23	-	12
Trait	27	-	21
Apathy Scale	5		
FrSBe- Self Total	63	-	8
FrSBe- Family Total	100	-	95

***Note:** Additional neuropsychological measures were given assessing all cognitive domains. Performances were within expectations.

Executive Functions		Raw	T-score	%ile
Set Shift & Prob Solve				
TMT-Part B		67	47	37
Wisconsin	# Categories	6		<16
	Persev Respon	16	41	18
	Persev Errors	16	40	14
	Total Errors	33	39	13

Cognitive Inhibition	Raw	T-score	%ile
Stroop Color-Word Trial	28	38	12
Stroop Interference Score	-4.7	45	30

Reward Sensitivity	Raw	T-Score	%ile
Iowa Gambling Task			
Deck A		-	6--10
Deck B, C, D		-	>16
Total Money	-1460		

***Figure 1.* Sagittal T3 weighted MR image. Arrows indicates displaced vermis**

Figure 2. Axial FLAIR MR image. Arrow indicates decreased vermis size.

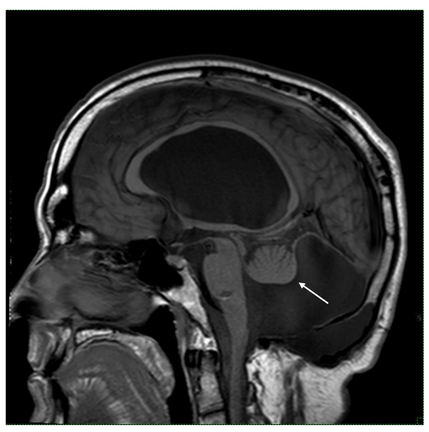


Figure 1

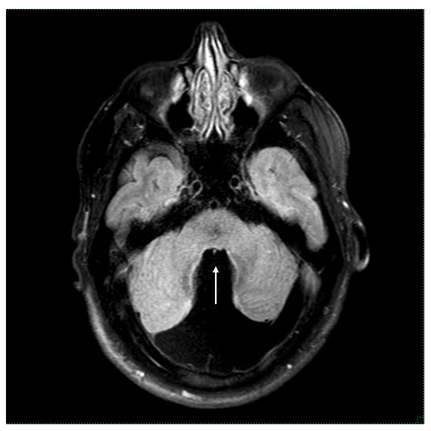
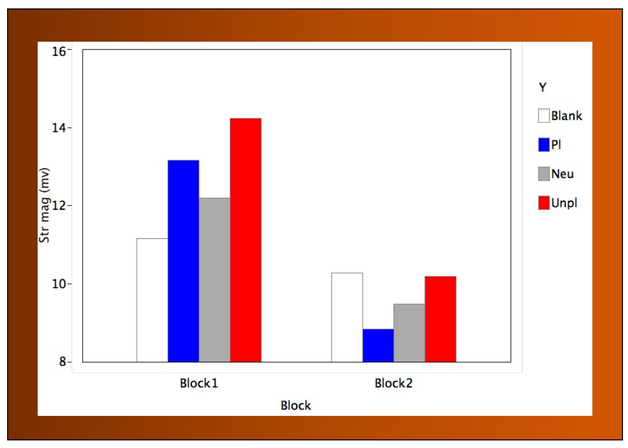


Figure 2

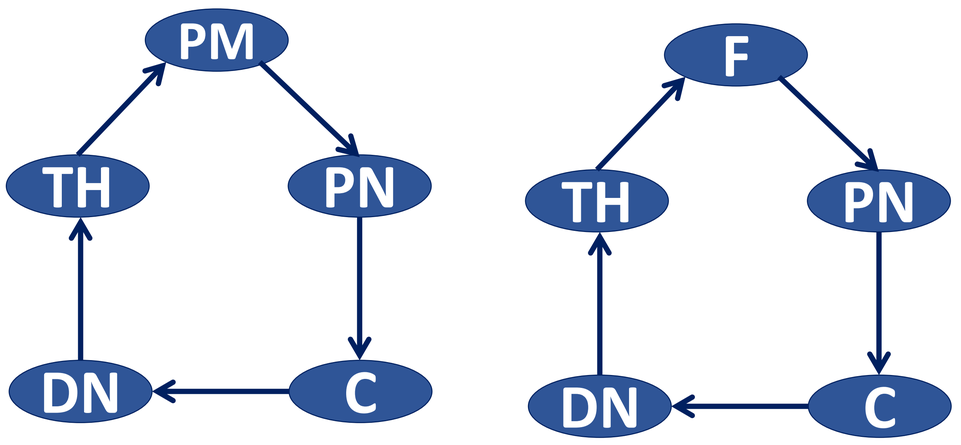
Magnitude of Startle Eyeblink while Dandy Walker patient viewed Pleasant, Unpleasant, & Neutral Pictures



No significant emotion modulatio n of startle magnitude. However, there was significant habituation of startle magnitude from Block 1 to Block 2 trials

Discussion

- More difficulties noted on experimental fronto-executive tasks associated with orbitofrontal- limbic circuitry
  - Reward insensitivity, mild physiological blunting
  - May indicate alteration in cerebellar-limbic- frontal network and represents aspects of CCAS
- Objective findings are consistent with functional concerns
  - **Clinical recommendations**: training with a motivational therapist to assist with insight and other related skills to improve functioning
- Findings indicate the need to expand testing beyond conventional neuropsychological measures
  - The use of atypical & experimental measures provided objective findings consistent with subjective report and functional neuroanatomical correlates
- **Limitations of study:** the constrained nature of testing may not fully capture the neurobehavioral weaknesses that are observed in daily life



Two examples of closed-circuit loops connecting the cerebellum with the cortex. *C= cerebellum, DN= dentate gyrus, TH= Thalamus, PM= Primary Motor Cortex, F= Dorsolateral Prefrontal Cortex* [6]

Works Cited: [1] Boddaert, N., Klein, O., Ferguson, N., Sonigo, P., Parisot, D. (2003) Intellectual prognosis of the Dandy- Walker malformation in children: The importance of vermian lobulation *Neuroradiology 45*(5), 320-340. . [2] Steinlin, M. (2008). Cerebellar disorders in childhood: Cognitive problems. *Cerebellum 7*(4), 607-610. [3] Schmahmann, J.D. (2010). The role of the cerebellum in cognition and emotion: Personal reflections since 1982 on the dysmetria of thought hypothesis, and its historical evolution from theory to therapy. *Neuropsychology Review., 20*(3), 236-260 . [4]Schmahmann, J.D. From movement to thought: Anatomic substrates of the cerebellar contribution to cognitive processing. *Human brain mapping, 4*(3), 174-198. [5]Alexiou, G.A., Sfakianos, G., & Prodromou, N. (2010). Dandy- Walker Malformation: Analysis of 19 cases. *Journal of Child Neurology 25*(2), 188-191. [6] Strick, P., Dum, R., & Fiex, J. (2009). *Cerebellum and nonmotor function*. Annual review of neuroscience, *32*, 413-434.