Prevalence Estimates For Three Types of Motor Speech Disorders In Complex Neurodevelopmental Disorders (CND)

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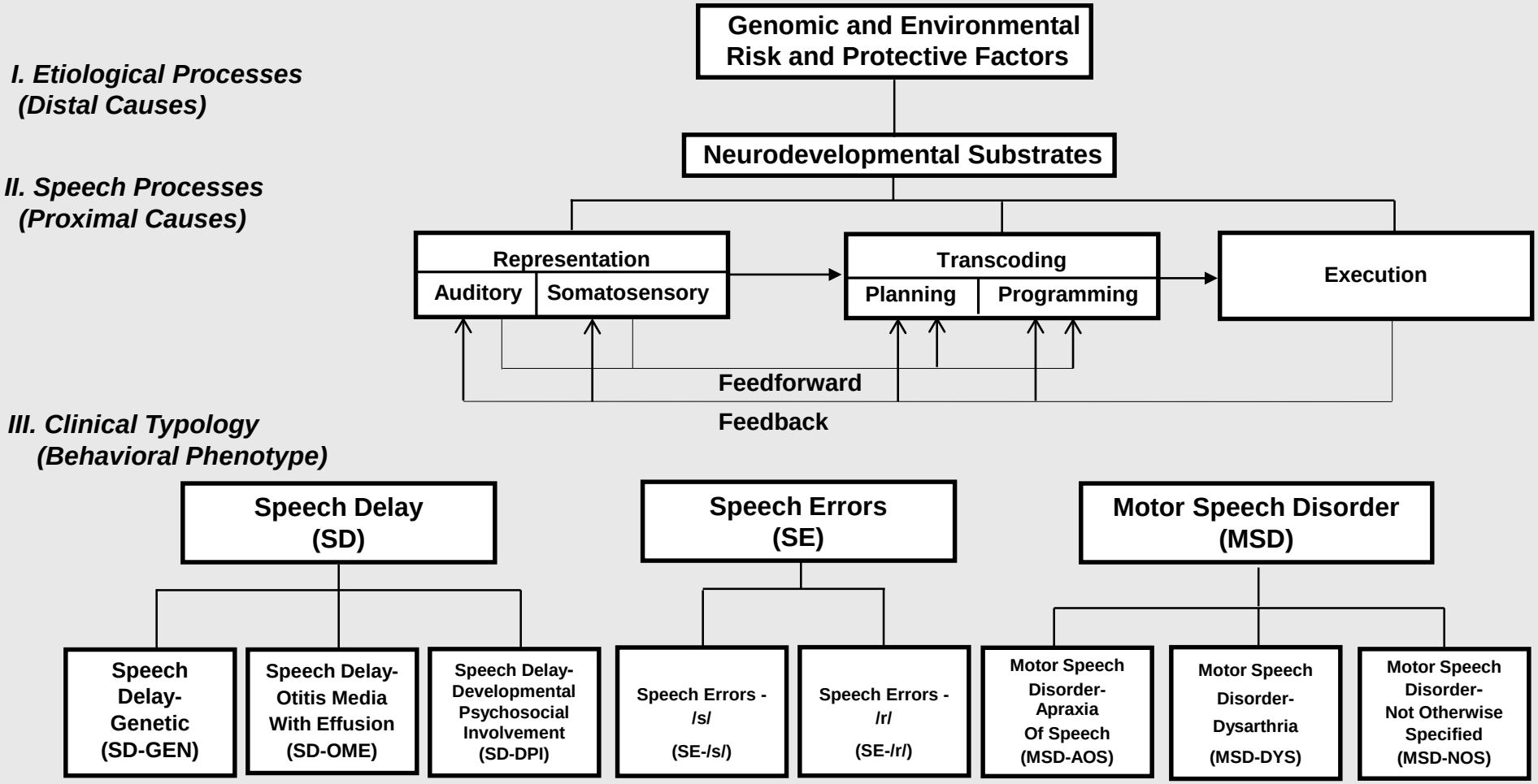
Statement of Purpose

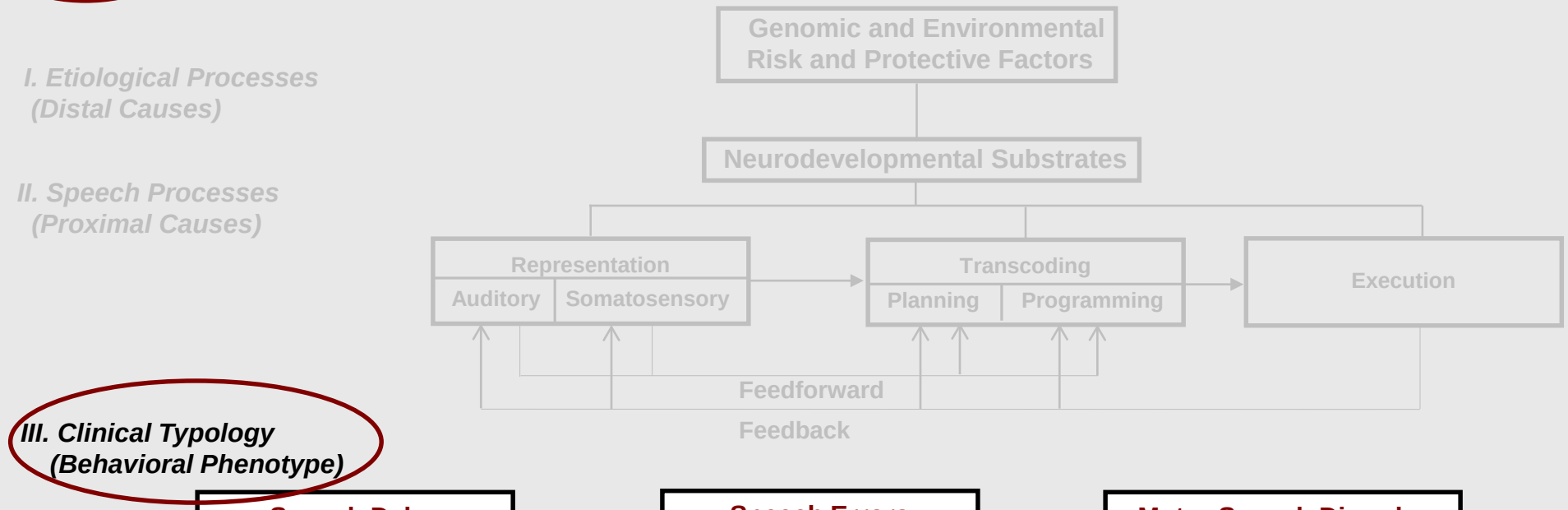
- Advances in the neurogenetics, treatment, and prevention of pediatric speech sound disorders require a nosology that cross-classifies a speaker's descriptive linguistic and motor speech status.
- Extensions to the finalized version of the Speech Disorders Classification System (SDCS) address this need.
- We summarize constructs, measures and prevalence estimates for three types of pediatric motor speech disorders in complex neurodevelopmental contexts.

Speech Disorders Classification System (SDCS)[a]



[a]Shriberg, L. D. (February, 2013). *State of the Art in CAS Diagnostic Marker Research.* Review paper presented at the Childhood Apraxia of Speech Association of North America Speech Research Symposium, Atlanta, GA.

Linear Representation of Speech and Motor Speech Classifications



III. Clinical Typology (Behavioral Phenotype)

- **Speech Delay (SD)**
  - Speech Delay-Genetic (SD-GEN)
  - Speech Delay-Otitis Media With Effusion (SD-OME)
  - Speech Delay-Developmental Psychosocial Involvement (SD-DPI)
- **Speech Errors (SE)**
  - Speech Errors - /s/ (SE-/s/)
  - Speech Errors - /r/ (SE-/r/)
- **Motor Speech Disorder (MSD)**
  - Motor Speech Disorder-Apraxia Of Speech (MSD-AOS)
  - Motor Speech Disorder-Dysarthria (MSD-DYS)
  - Motor Speech Disorder-Not Otherwise Specified (MSD-NOS)

IV. Diagnostic Markers (Criterial Signs of Phenotype)

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- - - - -
- - - - -

[a]Shriberg, L. D. (February, 2013). *State of the Art in CAS Diagnostic Marker Research.* Review paper presented at the Childhood Apraxia of Speech Association of North America Speech Research Symposium, Atlanta, GA.

Cross-Classification (Matrix) Representation of Speech and Motor Speech Classifications

SDCS SUMMARY I: SPEECH DISORDER AND MOTOR SPEECH DISORDER CLASSIFICATIONS								
Speeech Classification		Motor Speech Classification					Totals	
		No Motor Speech Disorder (NO MSD)	Motor Speech Disorder-Not Otherwise Specified (MSD-NOS)	Motor Speech Disorder-Dysarthria (MSD-DYS)	Motor Speech Disorder-Childhood Apraxia of Speech (MSD-CAS)	Motor Speech Disorder-Dysarthria and Childhood Apraxia of Speech (MSD-DYS & CAS)	n	%
Normal(ized) Speech Aquisition (NSA)								
Speech Errors (SE) Persistent Speech Errors (PSE) (SE/PSE)		____	____	____	____	____	____	____
Speech Delay (SD) Persistent Speech Delay (PSD) (SD/PSD)		____	____	____	____	____	____	____
Totals	n							
	%							

Cross-Classification (Matrix) Representation of Speech and Motor Speech Classifications

SDCS SUMMARY I:
SPEECH DISORDER AND MOTOR SPEECH DISORDER CLASSIFICATIONS

Speeech Classification		Motor Speech Classification					Totals	
		No Motor Speech Disorder (NO MSD)	Motor Speech Disorder-Not Otherwise Specified (MSD-NOS)	Motor Speech Disorder-Dysarthria (MSD-DYS)	Motor Speech Disorder-Childhood Apraxia of Speech (MSD-CAS)	Motor Speech Disorder-Dysarthria and Childhood Apraxia of Speech (MSD-DYS & CAS)	n	%
Normal(ized) Speech Aquisition (NSA)								
Speech Errors (SE) Persistent Speech Errors (PSE) (SE/PSE)		____	____	____	____	____	____	____
Speech Delay (SD) Persistent Speech Delay (PSD) (SD/PSD)		____	____	____	____	____	____	____
Totals	n							
	%							

Cross-Classification (Matrix) Representation of Speech and Motor Speech Classifications

SDCS SUMMARY I:
SPEECH DISORDER AND MOTOR SPEECH DISORDER CLASSIFICATIONS

Speeech Classification		Motor Speech Classification					Totals	
		No Motor Speech Disorder (NO MSD)	Motor Speech Disorder-Not Otherwise Specified (MSD-NOS)	Motor Speech Disorder-Dysarthria (MSD-DYS)	Motor Speech Disorder-Childhood Apraxia of Speech (MSD-CAS)	Motor Speech Disorder-Dysarthria and Childhood Apraxia of Speech (MSD-DYS & CAS)	n	%
Normal(ized) Speech Aquisition (NSA)								
Speech Errors (SE) Persistent Speech Errors (PSE) (SE/PSE)		____	____	____	____	____	____	____
Speech Delay (SD) Persistent Speech Delay (PSD) (SD/PSD)		____	____	____	____	____	____	____
Totals	n							
	%							

Cross-Classification of Speech and Motor Speech Disorders: Individual

SDCS SUMMARY I: SPEECH DISORDER AND MOTOR SPEECH DISORDER CLASSIFICATIONS								
Speeech Classification		Motor Speech Classification					Totals	
Participant PT03		No Motor Speech Disorder (NO MSD)	Motor Speech Disorder-Not Otherwise Specified (MSD-NOS)	Motor Speech Disorder-Dysarthria (MSD-DYS)	Motor Speech Disorder-Childhood Apraxia of Speech (MSD-CAS)	Motor Speech Disorder-Dysarthria and Childhood Apraxia of Speech (MSD-DYS & CAS)	n	%
Normal(ized) Speech Aquisition (NSA)			X					
Speech Errors (SE) Persistent Speech Errors (PSE) (SE/PSE)								
Speech Delay (SD) Persistent Speech Delay (PSD) (SD/PSD)								
Totals	n							
	%							

Cross-Classification of Speech and Motor Speech Disorders: Group[a]

SDCS SUMMARY I: SPEECH DISORDER AND MOTOR SPEECH DISORDER CLASSIFICATIONS								
Speeech Classification		Motor Speech Classification					Totals	
Down syndrome (*n*=50)		No Motor Speech Disorder (NO MSD)	Motor Speech Disorder-Not Otherwise Specified (MSD-NOS)	Motor Speech Disorder-Dysarthria (MSD-DYS)	Motor Speech Disorder-Childhood Apraxia of Speech (MSD-CAS)	Motor Speech Disorder-Dysarthria and Childhood Apraxia of Speech (MSD-DYS & CAS)	n	%
Normal(ized) Speech Aquisition (NSA)		0	0	1	0	0	1	2.0
Speech Errors (SE)		0	0	0	0	0	0	0.0
Persistent Speech Errors (PSE)		0	1	0	1	0	2	4.0
(SE/PSE)		0	1	0	1	0	2	4.0
Speech Delay (SD)		0	1	1	0	0	2	4.0
Persistent Speech Delay (PSD)		1	12	18	4	10	45	90.0
(SD/PSD)		1	13	19	4	10	47	94.0
Totals	n	1	14	20	5	10	50	
	%	2.0	28.0	40.0	10.0	20.0		100.0

[a] Wilson, E. M., Abbeduto, L., Camarata, S. M., & Shriberg, L. D. (2016). *Prevalence of speech and motor speech disorders and intelligibility correlates in persons with Down syndrome*. Manuscript in preparation.

Assessment

- Conversational speech task
- Syllable Repetition Task **(Shriberg & Lohmeier, 2008)**
- Data reduction
  - Narrow phonetic transcription
  - Prosody-voice coding
  - Acoustic analyses
- Data analyses
  - PEPPER **(Shriberg, Allen, McSweeny, & Wilson, 2001)**

Standardized Measures of Motor Speech Disorders

Motor Speech Classification	Measure	Description[a]
MSD-NOS	Precision-Stability Index (PSI)	32 speech, prosody, voice signs
MSD-CAS	Pause Marker (PM)	Four types of between-word inappropriate pauses
MSD-DYS	Dysarthria Index (DI) Dysarthria Subtype Indices (DSI)	34 speech, prosody, and voice signs 12-19 sign indices of five subtypes of dysarthria: Ataxic, Spastic, Hyperkinetic, Hypokinetic, Flaccid

[a] **Duffy (2013); Mabie & Shriberg (2016); Shriberg & Mabie (2016); Shriberg, Strand, Fourakis, Jakielski, Hall, Karlsson, Mabie, McSweeny, Tilkens, & Wilson (2016); Tilkens, Karlsson, Fourakis, Hall, Mabie, McSweeny, Wilson, & Shriberg (2016).**

Participants[a]

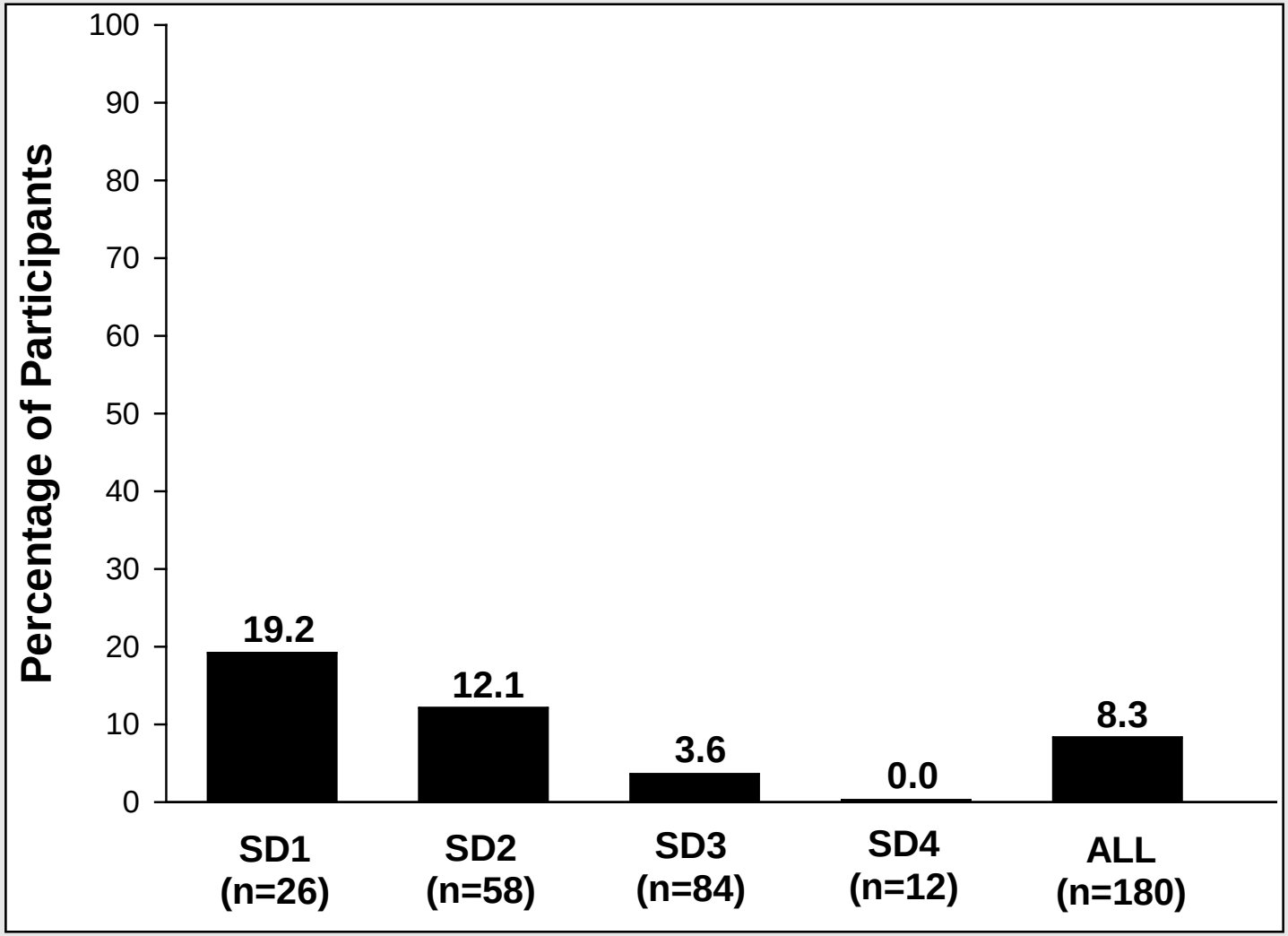
- CND Groups
  - 391 participants: 16p11.2, 22q11.2, ASD, CAS, DS, FXS, GALT, IID, TBI
- Typical-speaking Reference Groups
  - 150 participants, 3-18 years of age
  - 50 participants, 20-80 years of age
- Comparison Groups
  - 180 participants, 3-5 years of age, Idiopathic SSD
  - 22 participants, 45-84 years of age, AAS (stroke and PPAOS subgroups)

[a] 16p11.2: 16p11.2 microdeletion syndrome; 22q11.2: 22q11.2 deletion syndrome; AAS: Adult-onset Apraxia of Speech; ASD: Autism Spectrum Disorder; CAS: Childhood Apraxia of Speech; DS: Down syndrome; FXS: Fragile X syndrome; GALT: Galactosemia; IID: Idiopathic Intellectual Disability; PPAOS: Primary Progressive Apraxia of Speech; SSD: Speech Sound Disorders; TBI: Traumatic Brain Injury

MSD-NOS Prevalence Estimates

- 8% in children with Idiopathic SD
- 27% in 4 CND with high prevalence of MSD
  19% in 3 CND with low prevalence of MSD
- The persistent MSD phenotype in some speakers with prior CAS

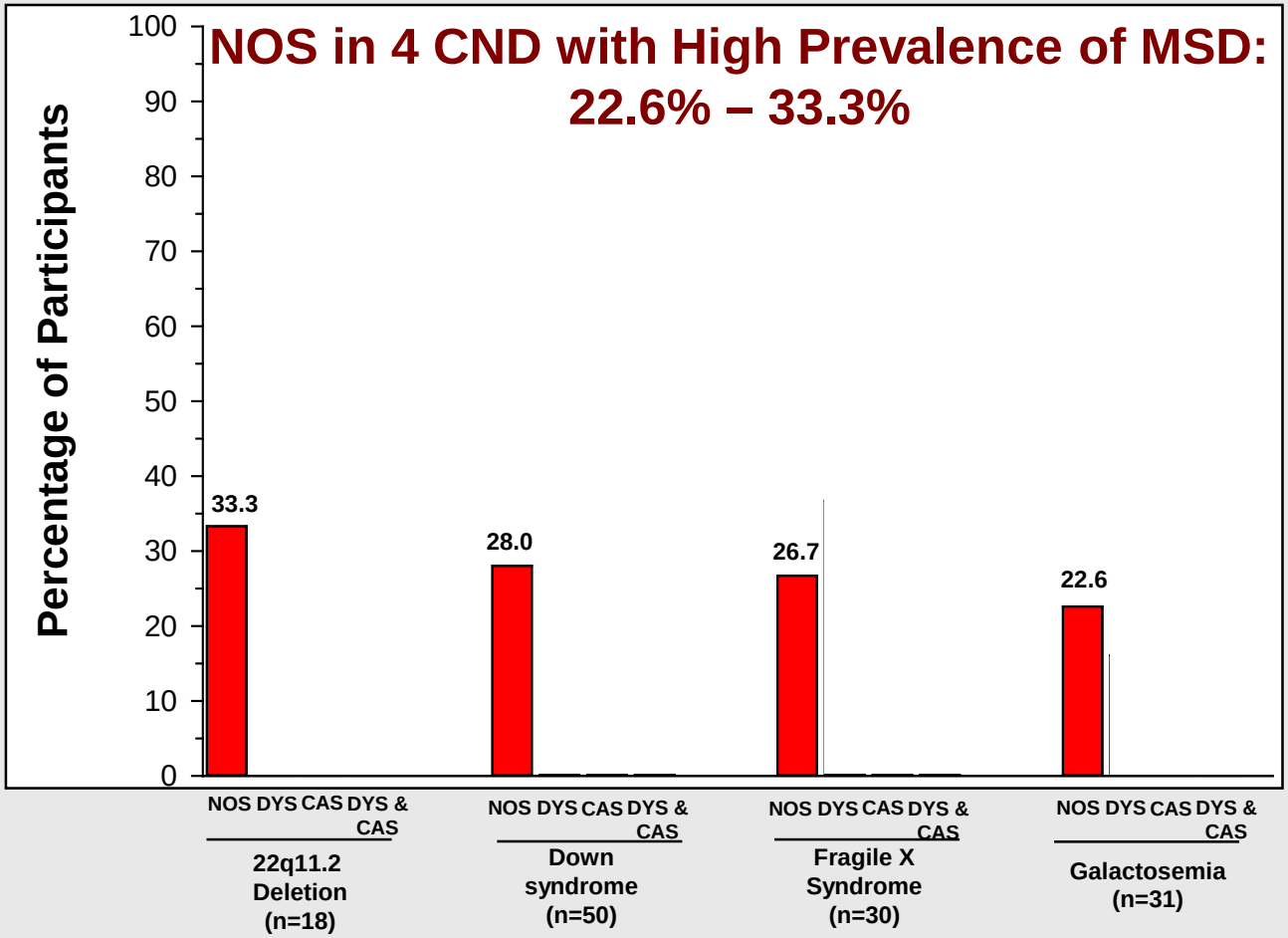
MSD-NOS Prevalence Estimates: In Idiopathic SD



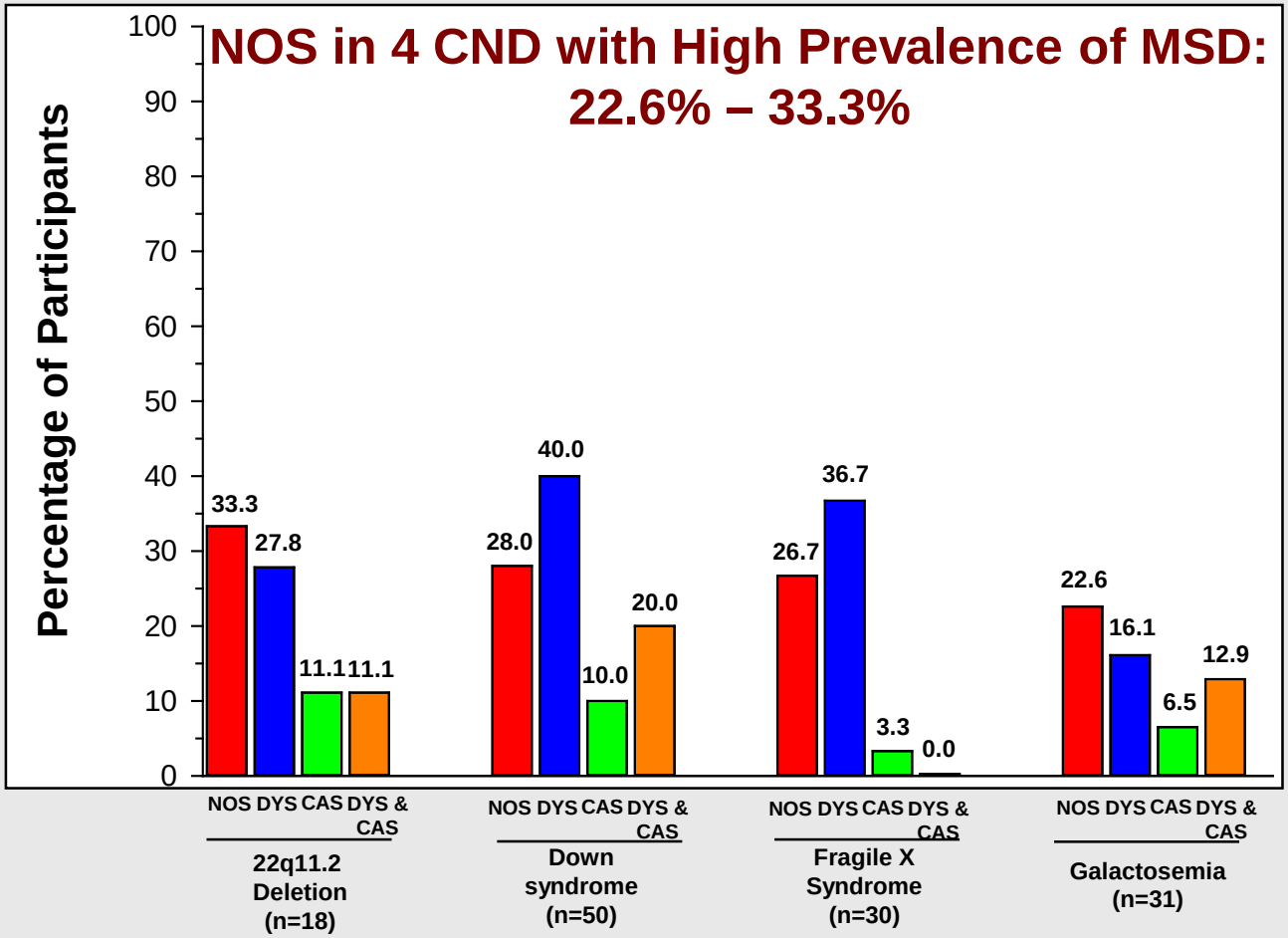
MSD-NOS Prevalence Estimates

- 8% in children with SD
- 27% in 4 CND with high prevalence of MSD
  19% in 3 CND with low prevalence of MSD
- The persistent MSD phenotype in some speakers with prior CAS

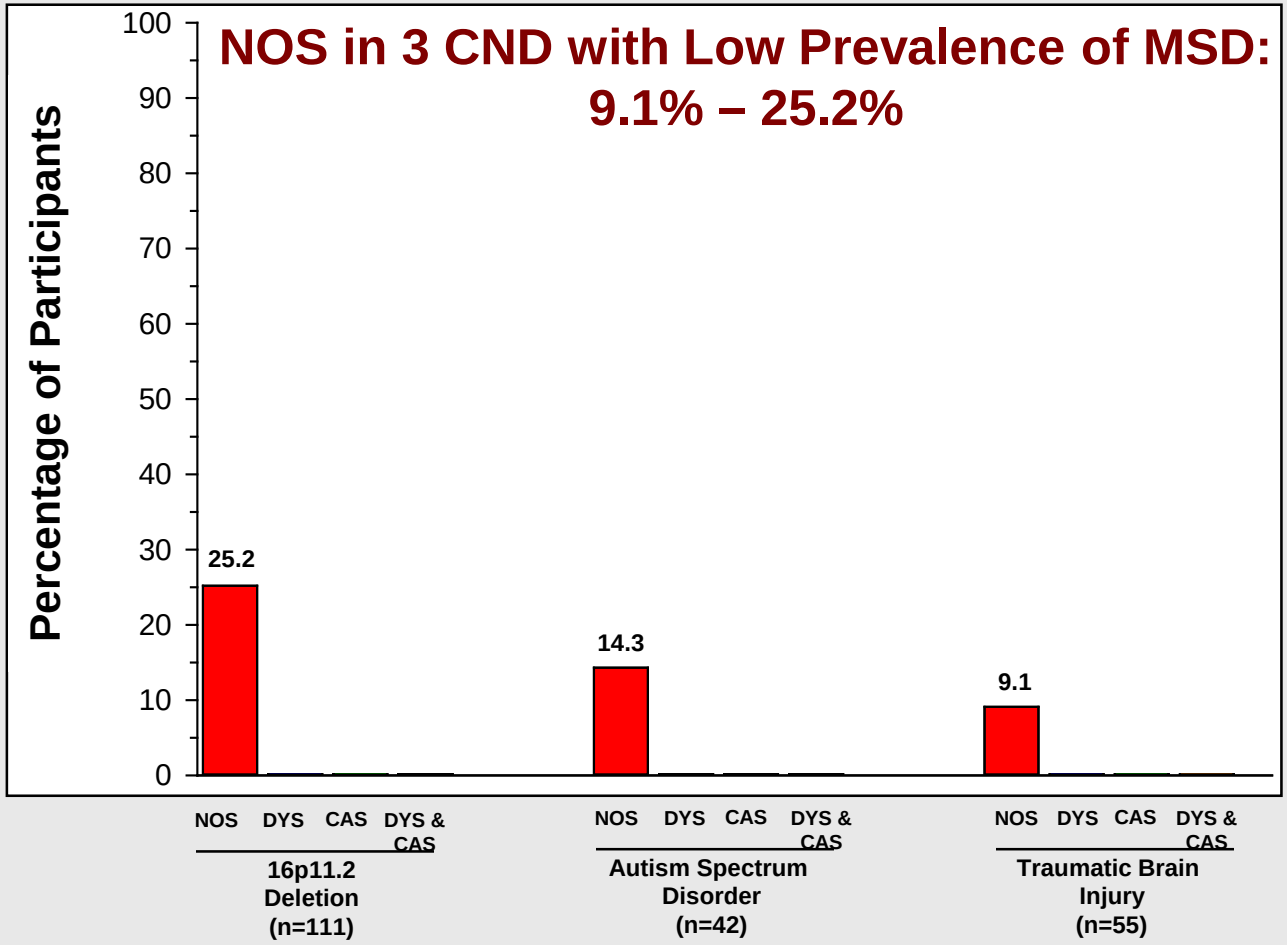
MSD-NOS Prevalence Estimates: In CND



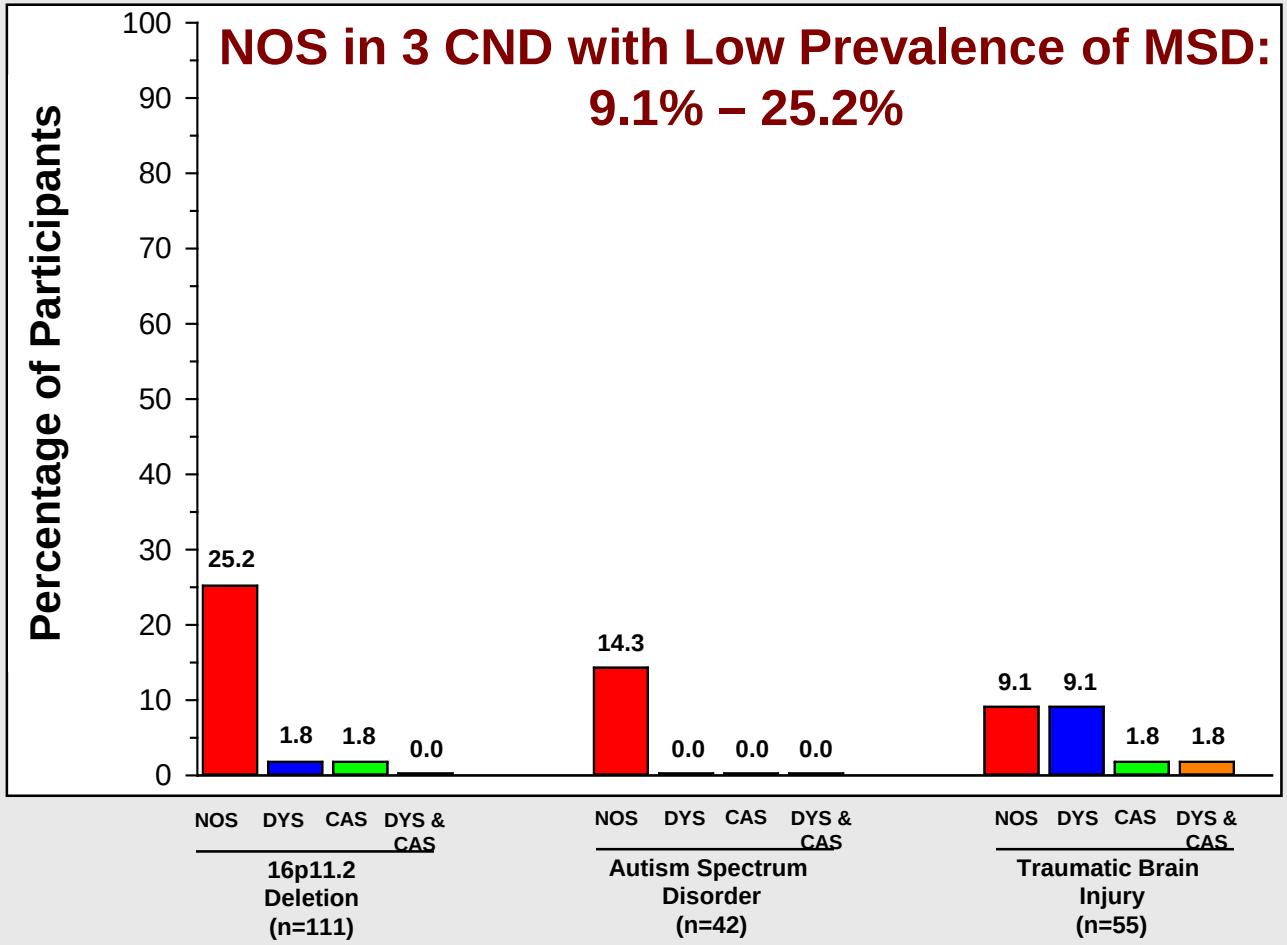
MSD-NOS Prevalence Estimates: In CND



MSD-NOS Prevalence Estimates: In CND



MSD-NOS Prevalence Estimates: In CND



MSD-NOS Prevalence Estimates

- 8% in children with SD
- 27% in 4 CND with high prevalence of MSD
  19% in 3 CND with low prevalence of MSD
- The persistent MSD phenotype in some speakers with prior CAS

MSD-NOS Prevalence Estimates: In Some Speakers with Prior CAS

SDCS SUMMARY I: SPEECH DISORDER AND MOTOR SPEECH DISORDER CLASSIFICATIONS								
Speeech Classification		Motor Speech Classification					Totals	
Patient 2[a]		No Motor Speech Disorder (NO MSD)	Motor Speech Disorder-Not Otherwise Specified (MSD-NOS)	Motor Speech Disorder-Dysarthria (MSD-DYS)	Motor Speech Disorder-Childhood Apraxia of Speech (MSD-CAS)	Motor Speech Disorder-Dysarthria and Childhood Apraxia of Speech (MSD-DYS & CAS)	n	%
Normal(ized) Speech Aquisition (NSA)								
Speech Errors (SE) Persistent Speech Errors (PSE) (SE/PSE)			X					
Speech Delay (SD) Persistent Speech Delay (PSD) (SD/PSD)								
Totals	n							
	%							

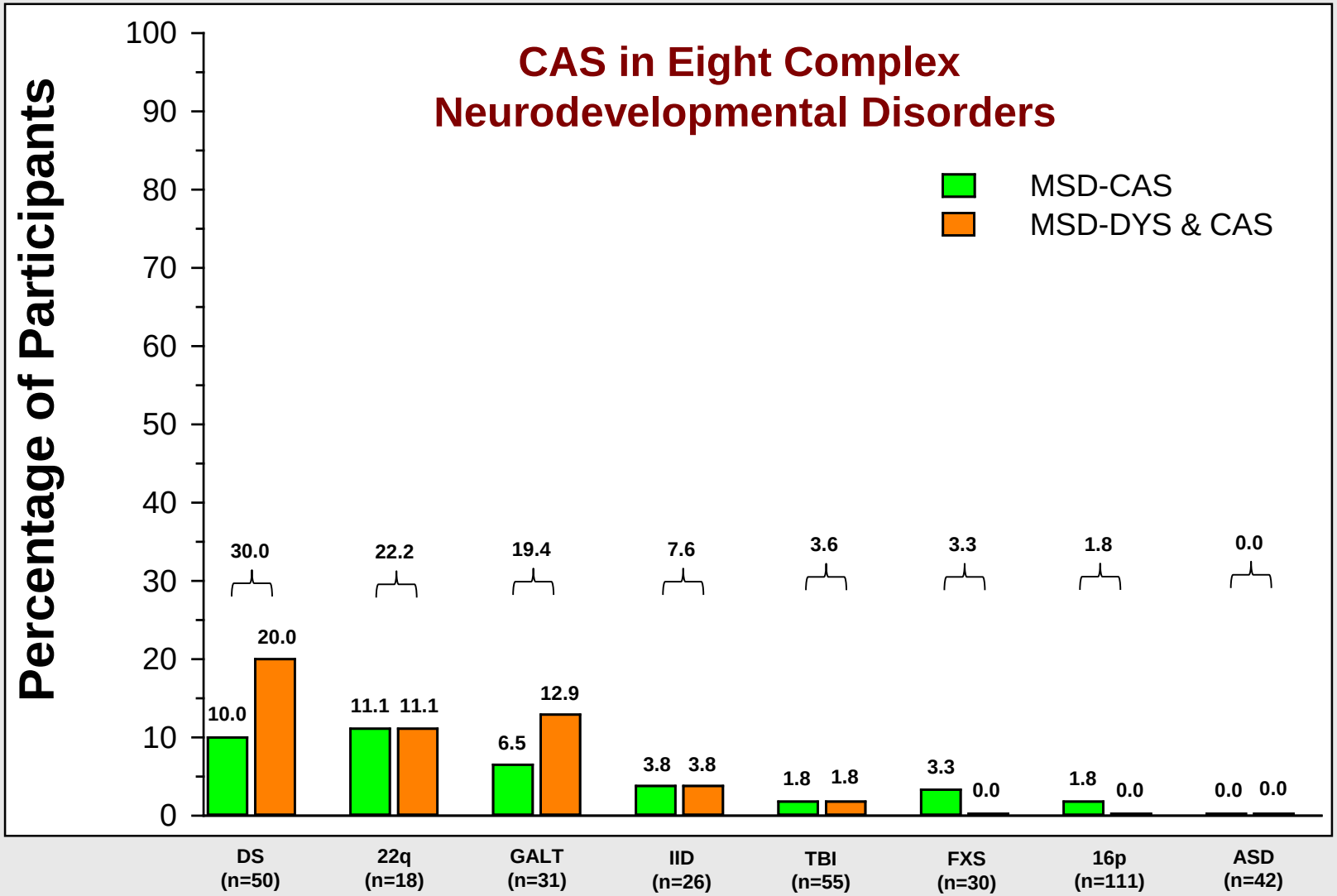
[a] Rice, G. M., Raca, G., Jakielski, K. J., Laffin, J. J., Iyama-Kurtycz, C., Hartley, S. L. . . Shriberg, L. D. (2012). Phenotype of *FOXP2* haploinsufficiency in a mother and son. *American Journal of Medical Genetics: Part A, 158A*, 174-181.

MSD-CAS & MSD-DYS & CAS Prevalence Estimates

- 0% - 30% CAS or DYS & CAS in 8 CND

- DYS & CAS more prevalent than CAS
  - in idiopathic CAS
  - in stroke and PPAOS forms of AAS
  - in several *FOXP2* case studies

MSD-CAS & MSD-DYS & CAS Prevalence Estimates

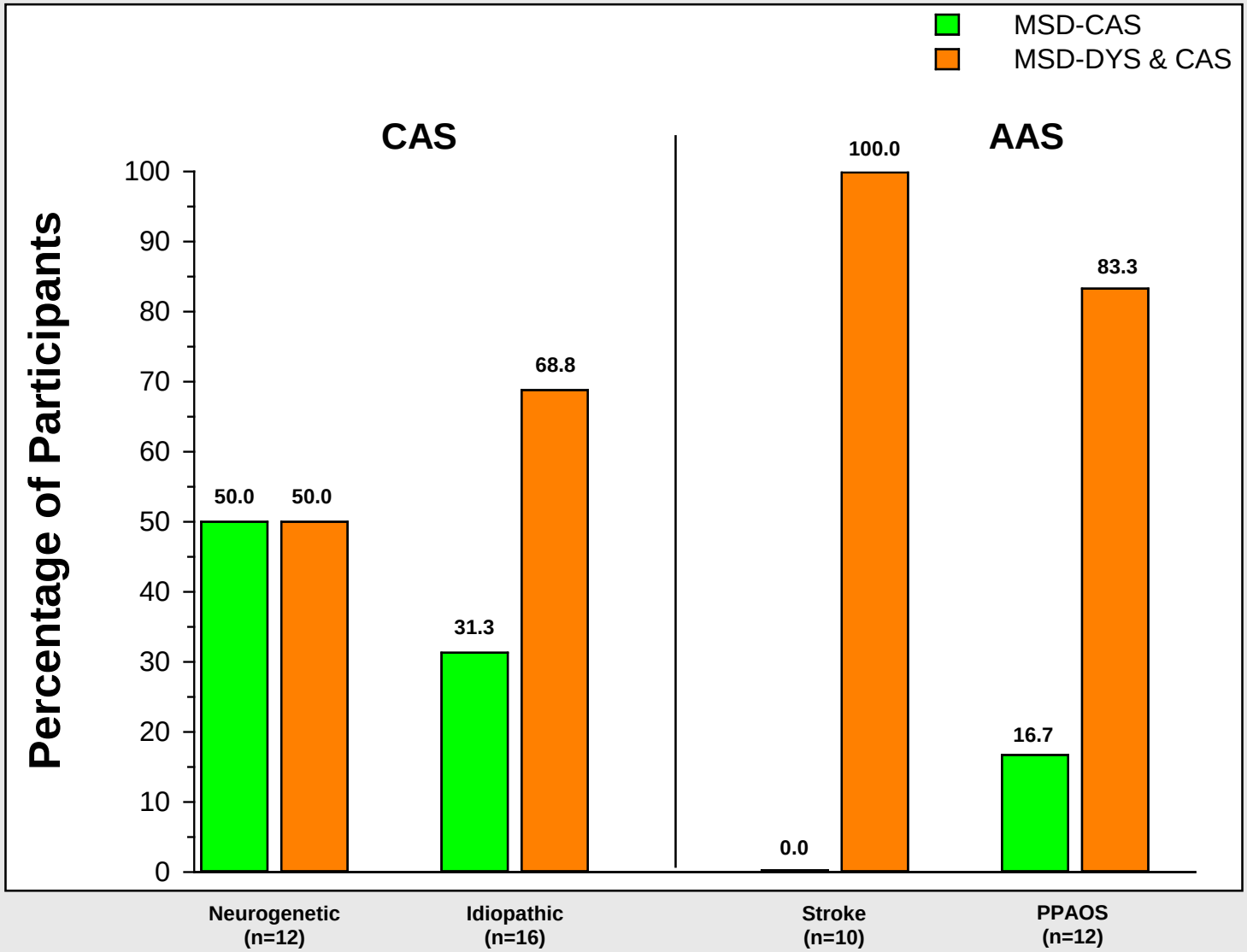


MSD-CAS & MSD-DYS & CAS Prevalence Estimates

- 0% - 30% CAS or DYS & CAS in 8 CND

- DYS & CAS more prevalent than CAS
  - in idiopathic CAS
  - in stroke and PPAOS forms of AAS
  - in several *FOXP2* case studies

MSD-CAS & MSD-DYS & CAS Prevalence Estimates



MSD-CAS & MSD-DYS & CAS Prevalence Estimates

- 0% to 39% CAS or DYS & CAS in 9 CND

- DYS & CAS more prevalent than CAS
  - in idiopathic CAS
  - in stroke and PPAOS forms of AAS
  - in several *FOXP2* case studies

MSD-CAS & MSD-DYS & CAS Prevalence Estimates

SDCS SUMMARY I: SPEECH DISORDER AND MOTOR SPEECH DISORDER CLASSIFICATIONS								
Speeech Classification		Motor Speech Classification					Totals	
Participant B (Shriberg et al., 2006; Tomblin et al., 2009)		No Motor Speech Disorder (NO MSD)	Motor Speech Disorder-Not Otherwise Specified (MSD-NOS)	Motor Speech Disorder-Dysarthria (MSD-DYS)	Motor Speech Disorder-Childhood Apraxia of Speech (MSD-CAS)	Motor Speech Disorder-Dysarthria and Childhood Apraxia of Speech (MSD-DYS & CAS)	n	%
Normal(ized) Speech Aquisition (NSA)								
Speech Errors (SE) Persistent Speech Errors (PSE) (SE/PSE)						X		
Speech Delay (SD) Persistent Speech Delay (PSD) (SD/PSD)								
Totals	n							
	%							

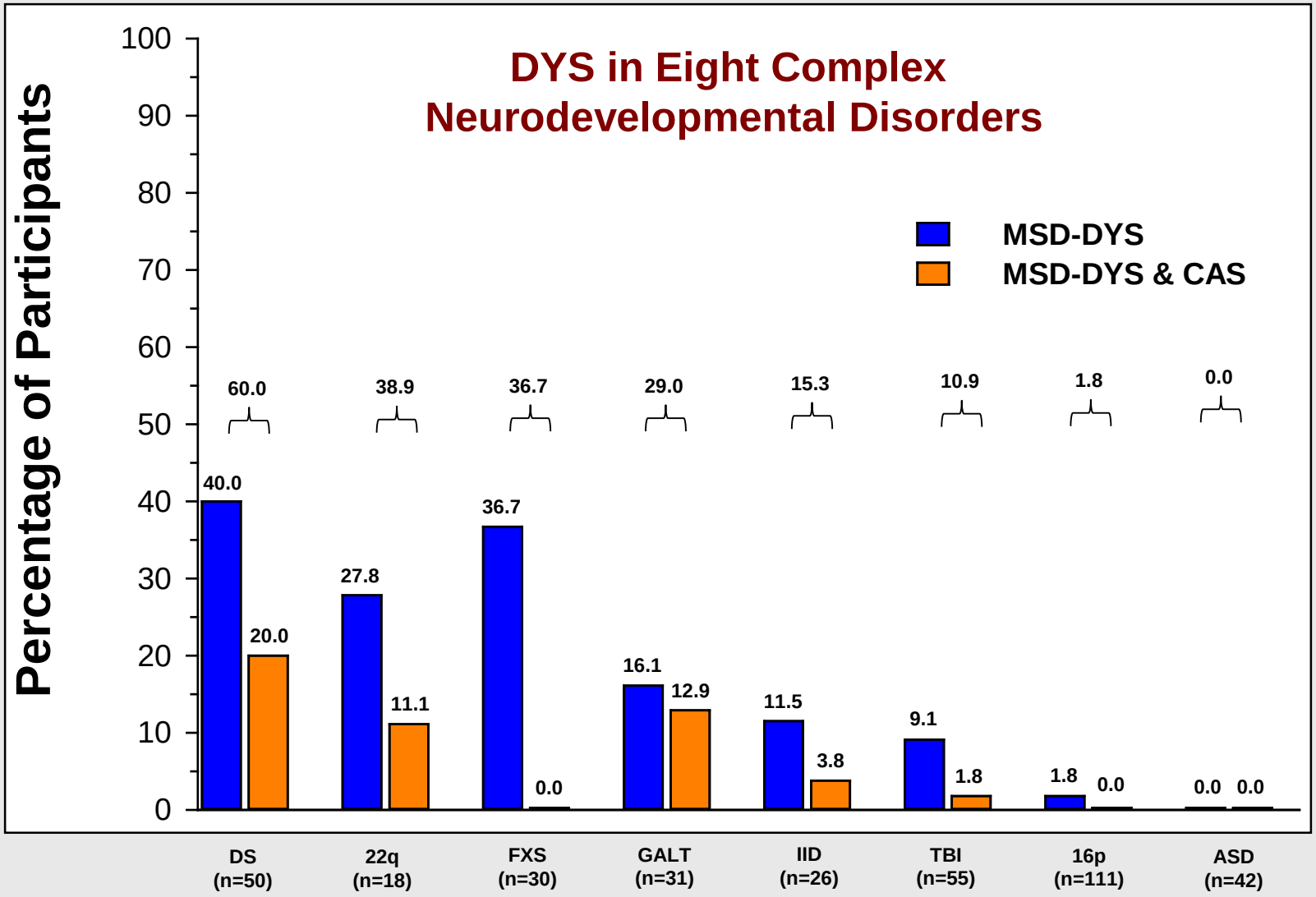
MSD-CAS & MSD-DYS & CAS Prevalence Estimates

SDCS SUMMARY I: SPEECH DISORDER AND MOTOR SPEECH DISORDER CLASSIFICATIONS								
Speeech Classification	Motor Speech Classification					Totals	
Participant T (Shriberg et al., 2006; Tomblin et al., 2009)	No Motor Speech Disorder (NO MSD)	Motor Speech Disorder-Not Otherwise Specified (MSD-NOS)	Motor Speech Disorder-Dysarthria (MSD-DYS)	Motor Speech Disorder-Childhood Apraxia of Speech (MSD-CAS)	Motor Speech Disorder-Dysarthria and Childhood Apraxia of Speech (MSD-DYS & CAS)	n	%
Normal(ized) Speech Aquisition (NSA)							
Speech Errors (SE) Persistent Speech Errors (PSE) (SE/PSE)							
Speech Delay (SD) Persistent Speech Delay (PSD) (SD/PSD)					X		
Totals	n						
	%						

MSD-DYS & MSD-DYS & CAS Prevalence Estimates

- 0% - 60% DYS or DYS & CAS in 8 CND
- DYS more prevalent than DYS & CAS in 7 CND
- Ataxic DYS most prevalent of the five dysarthria subtypes in CAS, AAS, DS, and FXS

MSD-DYS & MSD-DYS & CAS Prevalence Estimates



MSD-DYS & MSD-DYS & CAS Prevalence Estimates

- 0% to 60% DYS or DYS & CAS in 8 CND
- DYS more prevalent than DYS & CAS in 7 CND
- Ataxic DYS most prevalent of the five dysarthria subtypes in CAS, AAS, DS, and FXS

MSD-DYS & MSD-DYS & CAS Prevalence Estimates for Subtypes of Dysarthria

Group	Number Dysarthric[a]	Dysarthria Subtype Indices[b]				
		Ataxic	Spastic	Hyperkinetic	Hypokinetic	Flaccid
CAS (*n*=28)						
DYS&CAS	17	+				
AAS (*n*=22)						
DYS&CAS	20	+	+			
DS (*n*=50)						
DYS	20	++				
DYS&CAS	10	+		+		
FXS (*n*=30)						
DYS	11	+			+	
TBI (*n*=55)						
DYS	5		+			+
22q (*n*=18)						
DYS	5			+	+	+
GALT (*n*=31)						
DYS	5			+		
DYS&CAS	4		+	+		++

[a] Minimum number of participants required = 4

[b] ++< 10th percentile in 75-100% of participants

\+ < 10th percentile in 50-74.9% of participants

Summary and Conclusions

- One or more of three motor speech disorders (NOS, DYS, CAS) had substantial prevalence in the present samples of participants with complex neurodevelopmental disorders, and in participants with idiopathic Speech Delay.
- NOS provides a useful classification entity for research and clinical applications.
- CAS & DYS was more prevalent than CAS only, with implications for dimensional versus categorical nosologies in pediatric motor speech disorders.
- Cross-classification of speech and motor speech disorders should provide informative phenotypes for explanatory, treatment, and preventive research.

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