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#### Maternal Morbidity Outcomes in Idiopathic Moyamoya Syndrome in New York State

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## BACKGROUND

- Pregnancy is associated with an increased risk of stroke in young women
- Idiopathic moyamoya syndrome (IMMS) is a rare condition characterized by progressive narrowing of large cerebral arteries resulting in flimsy collaterals prone to rupture or thrombosis
- Data are limited on pregnancy outcomes in women with IMMS
- We hypothesized that IMMS would be associated with increased pregnancy morbidity, including stroke

#### METHODS

- Using the New York State Department of Health Statewide Planning and Research Cooperative System data from 2000-2014 and the International Classification for Diseases Ninth Edition (ICD-9), we identified all women aged 18 and older with diagnoses of IMMS (ICD-9 437.5) who had hospitalizations for delivery at any time either prior, concomitant or subsequent to IMMS diagnosis
- We excluded patients with Down syndrome (ICD-9 758.0), neurofibromatosis type 1 (ICD-9 237.71) and sickle cell disease (ICD-9 282.6) at time of IMMS diagnosis
- We then aggregated all pregnancies for these identified patients occurring between January 1, 1994 to December 31, 2014
  - Pregnancies were considered exposed if IMMS diagnosis occurred prior to or within 1 year of delivery
  - Intermediate unexposed pregnancies were those within 2-5 years prior IMMS diagnosis
  - Unexposed pregnancies occurred 6 or more years prior IMMS diagnosis
- Pregnancy morbidity was defined as admission within 1 year of delivery for any of the Center for Disease Control and Prevention's severe maternal morbidity indicators, including stroke
- We compared the morbidity of IMMS-exposed pregnancies to intermediate unexposed and unexposed pregnancies
- Generalized estimating equations were used to calculate odds ratio (OR) and 95% confidence intervals (95%CI) as well as adjust for women with multiple pregnancies occurring in both exposed and unexposed periods

### Maternal morbidity outcomes in idiopathic moyamoya syndrome in New York State Hajere J. Gatollari MPH, Amelia K. Boehme PhD MSPH, E. Sander Connolly MD, Alexander M. Friedman MD New York MPH, Mitchell S. V. Elkind MD MS MPhil, Joshua Z. Willey MD MS, Eliza C. Miller MD

#### Fig 1. Study Population Flow Diagram 852 IMMS patients from 2000-2014 626 MMS adults (18+) 554 idiopathic MMS adults (18+)386 IMMS women 134 IMMS parous women Table 1: Population demographics and characteristics Age (Mean + Standard deviation) Insurance Status Medicare/Medicaid Private Other **Pregnancy Number** 1 (First pregnancy) 2 (Second pregnancy) 3+ (Third or greater pregnancy) CDC indicator Stroke CDC indicator and/or stroke Death **ECIC** C section Table 2: Racial distribution of IMMS parous women Non-Hispanic White Non-Hispanic Black Hispanic Other Asian

# RESULTS

RESULTS Fig 2. Distribution of exposed and We identified 134 patients with unexposed pregnancies 264 pregnancies in total • A majority of pregnancies were unexposed (45.1%, n=119) compared to exposed (31.4%, <u>Both</u> <u>Unexposed</u> n= 83) and intermediate Exposed 23 patients 83 patients 28 patients 80 pregnancies unexposed (23.4%, n=62) 145 pregnancies 39 pregnancies -36 unexposed • There were 23 (17.1%) women -44 exposed that had both exposed and unexposed pregnancies • Severe maternal morbidity was highest for exposed pregnancies compared to intermediate unexposed and unexposed (34.9% vs. 27.4% vs 17.7%; p=0.0003) value After adjusting for age and multiple pregnancies, there were no significant odds of severe maternal morbidity or stroke in exposed pregnancies compared 0.01 to unexposed pregnancies (OR: 2.3, 95% CI: 0.8-6.9) CONCLUSION .0001 Pregnancies within 1 year prior .001 or any time after IMMS 0.02 diagnosis did not have .0003 increased maternal morbidity 0.3 compared to unexposed .0001 pregnancies after adjusting for .0001 age and clustering of women with multiple pregnancies **P-value** 

0.1

0.4

- Excluded: 226 children (<18 years old)
  - Excluded: 72 secondary causes Sickle cell disease (n=60) Down Syndrome (n=9) Neurofibromatosis Type 1 (n=3)
    - Excluded: 168 men

Excluded: 252 nulliparous women

Exposed	(n=83)	Intermediate Unexposed (n=62)	Unexposed (n=119)	P-v	
N (%)		N (%)	N (%)		
26.2 <u>+</u> 7.3		32.2 <u>+</u> 7.3	39.0 <u>+</u> 8.0		
21 (25	.3%)	15 (24.2%)	19 (16.0%)		
11 (13	.3%)	1 (1.6%)	6 (5.0%)		
51 (61	.4%)	46 (74.2%)	94 (79.0%)		
28 (20	.9%)	35 (26.1%)	71 (53.0%)	<.(	
21 (33.9%)		12 (19.4%)	29 (46.8%)		
34 (50.0%)		15 (22.1%)	19 (27.9%)		
25 (30.1%)		15 (24.2%)	19 (16.0%)	0.	
6 (7.2%)		2 (3.2%)	3 (2.5%)	0	
29 (34.9%)		17 (27.4%)	21 (17.7%)	0.0	
1 (1.2%)		0 (0.0%)	0 (0.0%)	(	
10 (12.1%)		0 (0.0%)	0 (0.0%)	<.(	
38 (45.8%)		27 (43.6%)	9 (7.6%)	<.(	
N (%)					

	Table 3: Adjusted GEE models	OR (95% CI)
51 (38.1%)	Age, CDC and stroke (ref = unexposed)	
27(20.1%)	Exposed	2.3 (0.8-6.9)
19 (14.2%)		<b>X 7</b>
24 (17.9%)	Intermediate unexposed	1.6 (0.6-4.1)
13 (9.7%)		

• Prospective studies are needed to better characterize increased maternal risks for women with moyamoya syndrome and develop preventive strategies