

# Facial Weakness and Impact on Communication and Swallowing in FSHD

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# My Objective

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- To evaluate a quantitative measure of facial weakness
- To determine the impact of FSHD on communication and swallowing difficulty.
- To determine how our quantitative measures of facial weakness compare to other aspects of FSHD (duration, severity, communication, swallowing difficulty, and physical function)

# Background

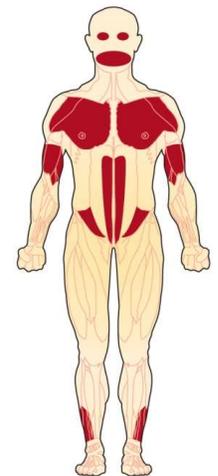
- FSHD is one of the most common muscular dystrophies
- 1 in 8,333 to 1 in 20,000
  - About 21,000 cases in USA
- Progression: face/scapular muscles, upper arms, lower legs/abdominal



Scapular winging due to asymmetric dystrophy of scapular muscles  
(<https://www.fshsociety.org>)



Facial weakness



(<https://www.mda.org>)

# Facioscapulohumeral Muscular Dystrophy

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- There is considerable variability between individuals but weakness is progressive over time
  - About 20% of individuals over the age of 50 will require a wheelchair
- Prior studies have shown a consistent loss of strength using manual muscle testing or quantitative myometry of about 3-5% per year
  - But there are no validated measures facial weakness
- Only limited studies regarding the impact of facial weakness on communication or swallowing

# Methods

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- Prospective, cross-sectional observational study
- Performed at two sites (KUMC/University of Utah)
- Inclusion criteria
  - > 18 years
  - Genetic confirmation (or immediate relative)
  - Clinically affected
  - Could complete study procedure
- Single-visit

# Methods

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- IOPI (Iowa Oral Performance Instrument)
  - Quantifies lip, tongue, and cheek pressure
- SWAL-QOL
  - Patient-reported swallowing difficulty
- CPIB (Communicative Participation Item Bank)
  - Patient-reported communication questionnaire

# Methods: IOPI

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- Lip, tongue, and cheek strength
- Displacement of air pressure
- Three trials, maximum



(<http://iopimedical.com/>)

# Methods: SWAL-QOL

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- Questionnaire to determine quality of life as affected by swallowing difficulties
- 10 categories, scored as a percentage of normal
  - (100% = healthy)
  - Categories include: food selection; burden; mental health; social functioning; fear; eating duration; eating desire; communication; sleep; and fatigue

# Methods: Communicative Participation Item Bank – short version

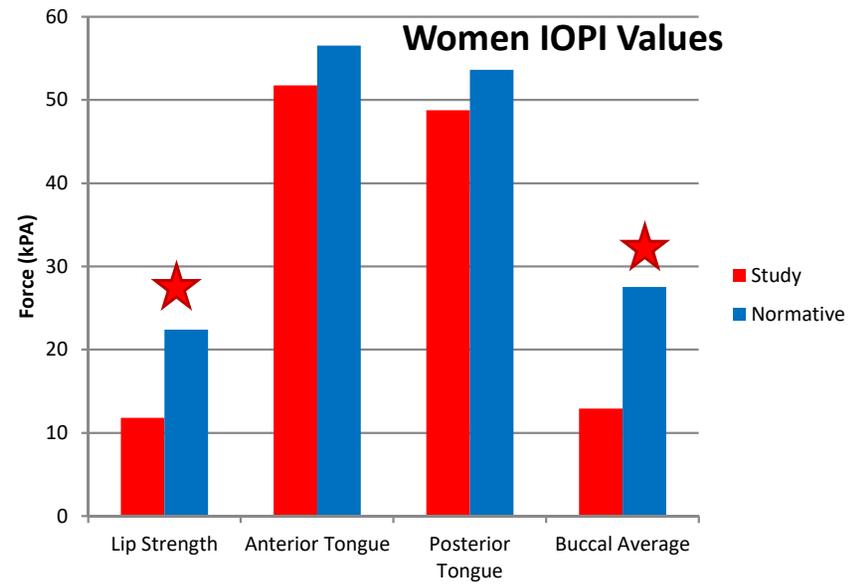
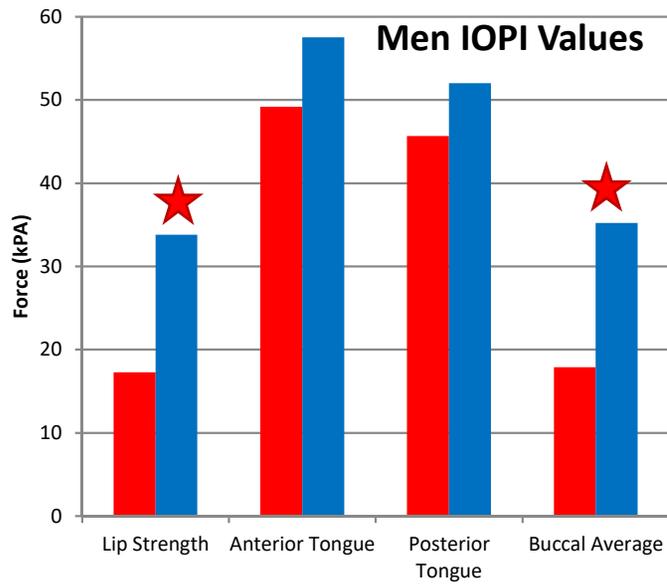
- 10-item questionnaire
  - Asks whether condition interferes with communication
  - Question: “Does your condition interfere with talking with people you know?”
  - Scored from 0=‘very affected’ to 3=‘not at all’
- Summary score out of 30

	Not at all	A little	Quite a bit	Very much
1. Does your condition interfere with.....talking with people you know?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
2. Does your condition interfere with.....communicating when you need to say something quickly?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
3. Does your condition interfere with.....talking with people you do NOT know?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
4. Does your condition interfere with.....communicating when you are out in your community (e.g. errands; appointments)?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
5. Does your condition interfere with.....asking questions in a conversation?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
6. Does your condition interfere with.....communicating in a small group of people?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
7. Does your condition interfere with.....having a long conversation with someone you know about a book, movie, show or sports event?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
8. Does your condition interfere with..... giving someone DETAILED information?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
9. Does your condition interfere with.....getting your turn in a fast-moving conversation?	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
10. Does your condition interfere with.....trying to persuade a friend or family member to see a different point of view	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

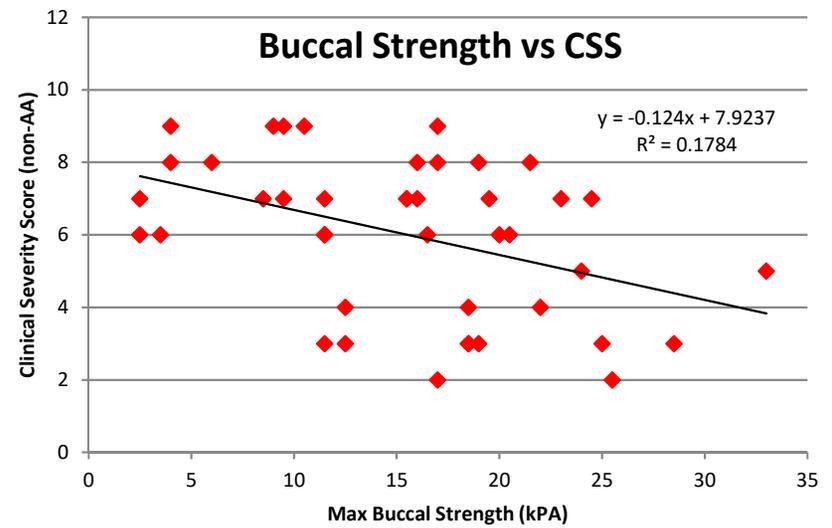
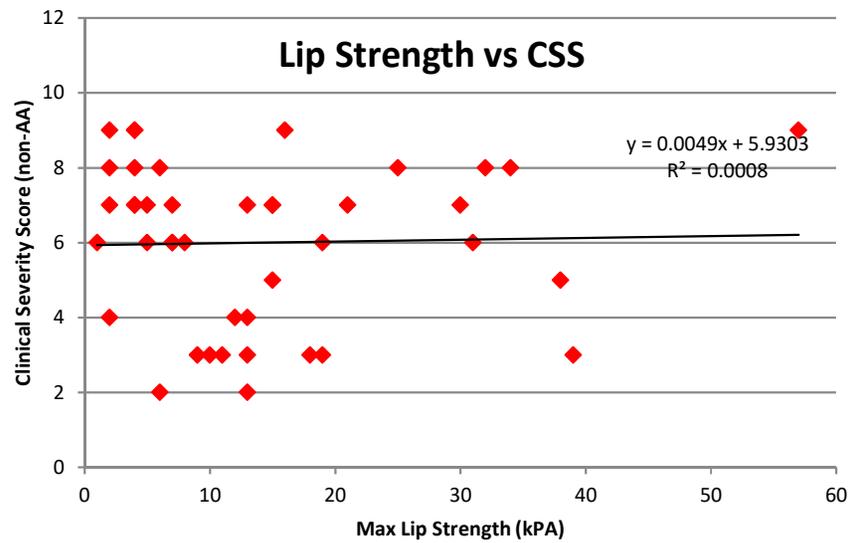
# Patient Demographics

	Kansas	Utah	Total (Range)
<b>Mean Age</b>	55.37	50.76	52.75 (23-83)
Range	23-83		
<b>Gender (% female)</b>	47.37%	44.00%	45.45%
<b>Mean Disease Duration (symptom onset)</b>	26.89	32.31	29.92
Range	5-64		
<b>Mean Clinical Severity Score (non age-adjusted)</b>	6.5	5.64	6
Range	2-9		
<b>FSHD Clinical Score</b>	9.055556	7.64	8.232558
Range	2-13		

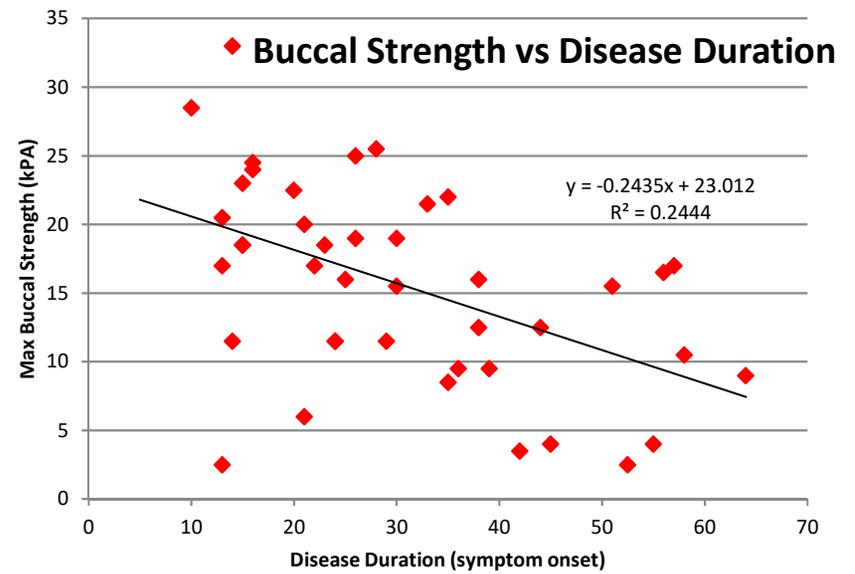
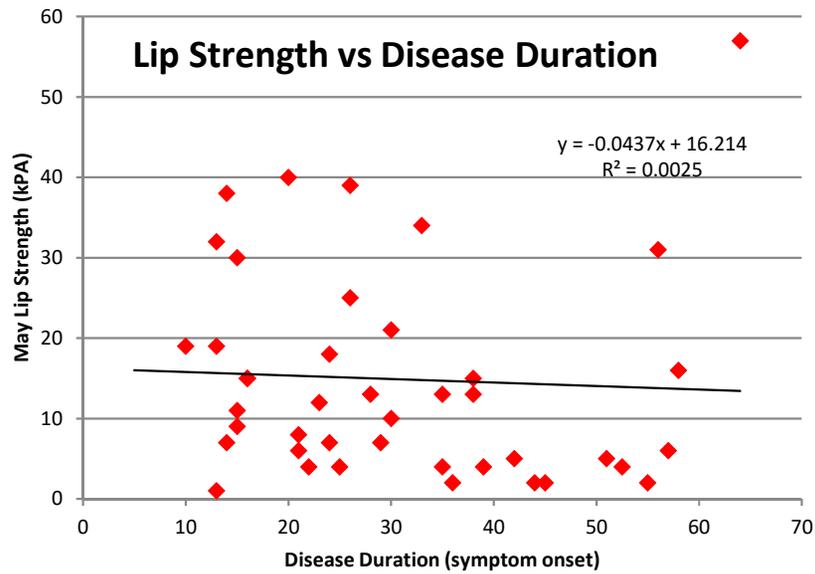
# IOPI



# Iowa Oral Pressure Instrument



# IOPI



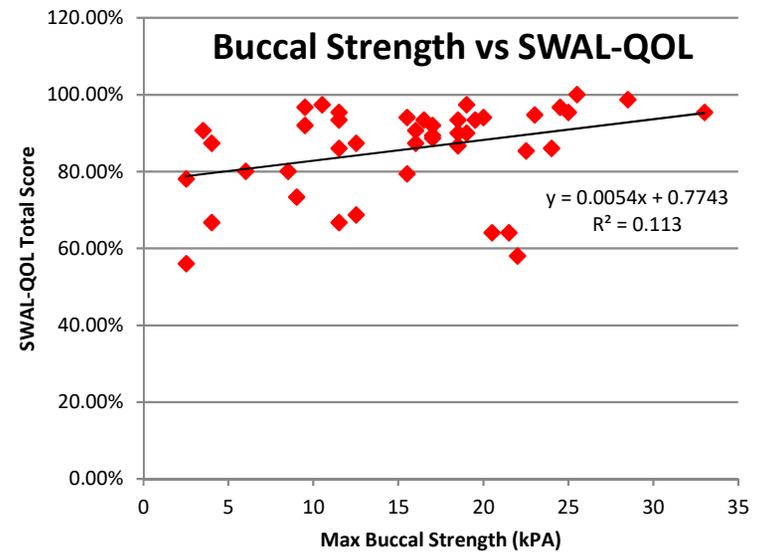
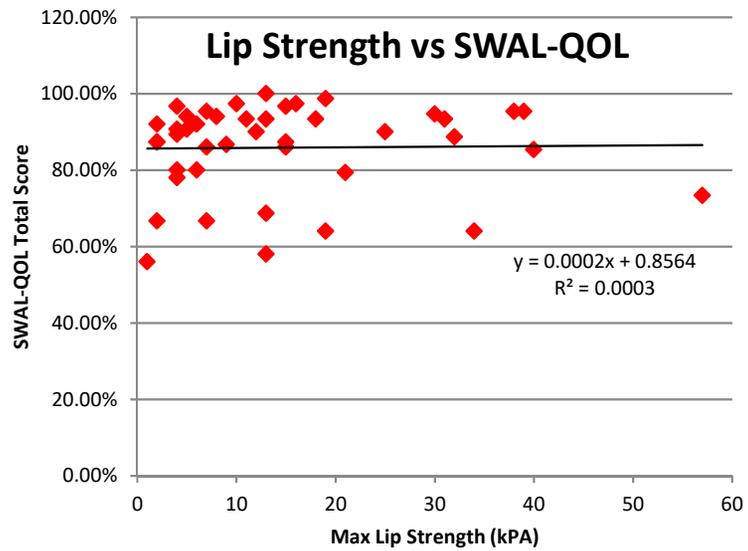
# SWAL-QOL

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	Mean	STD
Burden	91%	14%
Eating Duration	86%	19%
Eating Desire	92%	12%
Food Selection	91%	15%
Communication	91%	13%
Fear	89%	14%
Mental Health	91%	17%
Social	93%	15%
Sleep	<u>65%</u>	22%
Fatigue	<u>55%</u>	20%
<b>Total</b>	<b>86%</b>	<b>12%</b>

- While the overall score is not that low, about 1/3 of people have a total SWAL-QOL score below 80%

# SWAL-QOL



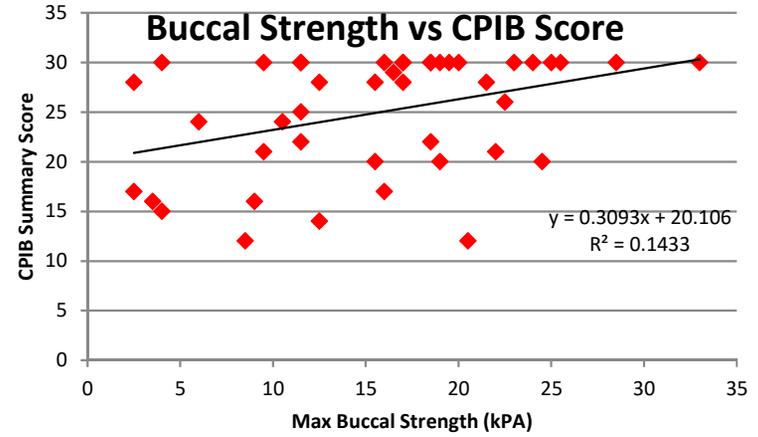
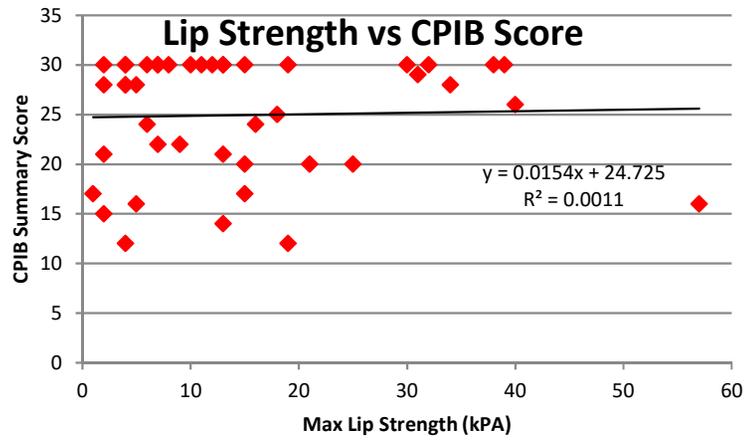
# CPIB

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Mean	24.84
Standard Deviation	5.93
N	44

- Ceiling affect with 18 reporting score of 30
- But the range as low as 12
- There was a positive correlation between the SWAL-QOL and the CPIB, with a  $r=0.61$

# CPIB



# Conclusion

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- The IOPI offers an easy to perform standard assessment face and tongue weakness for FSHD
  - Includes standard protocols and normative values
  - Both lip strength and buccal strength were reduced in FSHD
- Buccal strength as a better indicator of disease state
  - With modest associations to overall severity and disease duration
  - Modest associations with communication difficulties or swallowing
  - May be good measure to follow over time
- While most FSHD patients score ~normal for swallow and communication; a smaller group, around 1/3 may have difficulties

# Implications

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- What does it mean?
- Focus on buccal strength and buccal movement
  - We often think of measuring changes in orbicularis oris strength, and while this is affected, it may be static
  - Buccal strength is combination of muscles (orbicularis oris, buccinator, etc) which together may show some progression over time
- Follow-up studies needed
  - Other factors beside lip or buccal strength which may affect communication or swallow
  - Other aspects of lip weakness not explored here: e.g. the social impact of not having facial expression understood

# Thank You

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- The research team at KU
- The research team at University of Utah
- Karlien Mul, a summer research fellow at KU working in FSHD
- The KU summer learning experience
- The patient and family members with FSHD who participated in our study
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