

## **CHRD 2020: Abstract Submission Form**

**Submitter Name** 

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#### Title

Protein Origami: The Bowen-Conradi Syndrome Protein Emg1 is Unfolded

#### Background

Bowen-Conradi Syndrome (BCS) is a ribosomopathy (ribosome assembly disorder) present in the prairie Hutterite population and is characterized by severe development delay leading to death in early childhood. There are no treatments available for individuals with BCS; therefore further research needs to be conducted in order to better understand the disease and to propose possible treatments solutions.

#### Objective

The aim of this project is to test whether the BCS variant (D86G) of the ribosome assembly protein Emg1, is unfolded as seen by a trypsin sensitivity assay. If the Emg1-BCS variant causes localized unfolding or otherwise perturbs the structure of Emg1, we expect to see a difference in the proteolytic digestion pattern in the presence of trypsin.

#### Methods

Using a yeast model of BCS, whole cell lysates were prepared from both wild-type and BCS cells. For the trypsin sensitivity assay, the lysates were incubated with a time course of trypsin. Proteins were resolved by SDS-PAGE and trypsin digestion of Emg1 was monitored by western blotting.

#### Results

Using both the human Emg1 and its BCS variant (D86G) along with its yeast homolog and BCS equivalent (D90G), we observe that the wild-type Emg1 is very stable whereas the BCS variant is rapidly degraded in our trypsin sensitivity assay. This consistent difference in digestion pattern between the Emg1 wild-type and BCS variants supports the claim that the Emg1 BCS variant is unfolded in comparison to wild type.

#### Conclusion

From this data, we conclude that BCS is due in part to local or global protein unfolding which would

therefore alter Emg1's protein stability, protein-protein interacting partners, and function.

Theme: Basic Science

# Do you have a table/figure to upload? No

Are you willing to participate in Goodbear's Den? No

**Presenter Status:** Undergraduate Students

What was your role in the project? Perform Experiments

### Authors

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