Are all neurodegenerative diseases caused by faulty proteins?

The GLP aggregated and excerpted this blog/article to reflect the diversity of news, opinion and analysis.

A review published in *Science* has once again sparked one of the great debates in neuroscience – namely, are all neurodegenerative diseases prions?

Prions have always been shrouded in controversy. Stanley Prusiner, who won the 1997 Nobel prize in Physiology and Medicine for his discovery of the infectious proteins, fought through years of belittling cynics. Once the prion phenomenon was accepted, Britain was struck by BSE (bovine spongiform encephalopathy or mad cow disease) and a national epidemic was feared.

Prions are unlike any other infectious diseases. Simply put, they are misfolded proteins that confer their misfolded state onto other, normal, cellular proteins. These misfolded proteins then clump together to form aggregates that disrupt cellular functions like protein transport and respiration. Finally, the cell dies. The protein that misfolds in all known prion disease has come to be known prion protein, or PrP for short.

All common neurodegenerative diseases are associated with distinct proteins. What is surprising is that in all of these diseases their respective proteins misfold to form aggregates. If this sounds familiar to you, that's because it's incredibly similar to the way PrP causes neurodegeneration in prion diseases like Kuru.

But this in itself doesn't implicate the proteins as the causative agents. For many years these proteins were thought to be the result of the disease, not its cause. It was thought that various genetic or environmental factors caused the disease, and then as a result of the disease these proteins began to misfold and aggregate.

Read full, original post: What's In a Name? The Big Prion Debate