can be considered part of the FTLD clinical spectrum. Furthermore, patients may evolve from any of the major syndromes described above to have prominent features of another syndrome.

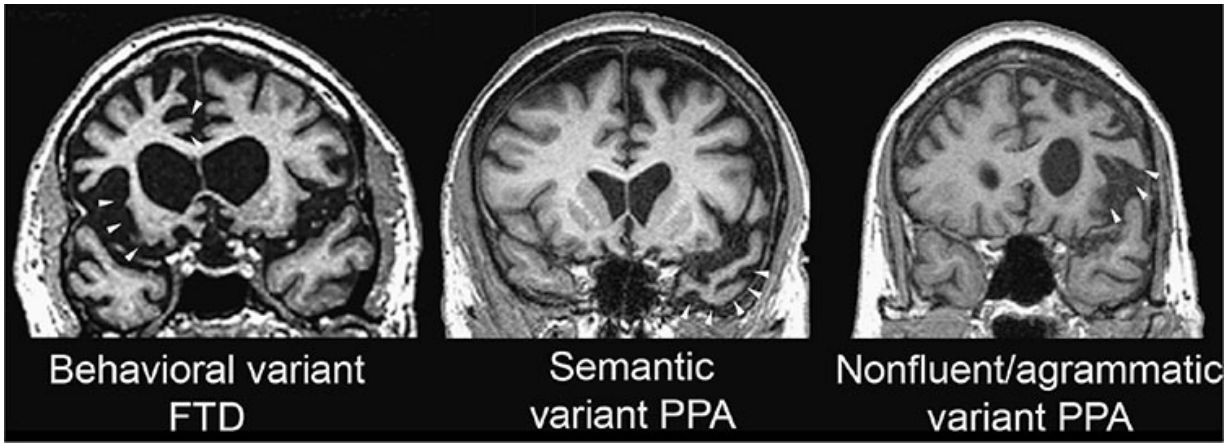

FIGURE 432-1 Three major frontotemporal dementia (FTD) clinical syndromes. Coronal MRI sections from representative patients with behavioral variant FTD (*left*) and the semantic (*center*) and nonfluent/agrammatic (*right*) variants of primary progressive aphasia (PPA). Areas of early and severe atrophy in each syndrome are highlighted (*white arrowheads*). The behavioral variant features anterior cingulate and frontoinsular atrophy, spreading to orbital and dorsolateral prefrontal cortex. Semantic variant PPA shows prominent temporopolar atrophy, more often on the left. Nonfluent/agrammatic variant PPA is associated with dominant frontal opercular and dorsal insula degeneration.

Findings at the bedside are dictated by the anatomic localization of the disorder. Degeneration with atrophy occurs in the medial and orbital frontal and anterior insula in bvFTD; the anterior temporal region in semantic variant PPA; and the lateral frontal and precentral gyrus of the dominant hemisphere in nonfluent/agrammatic PPA. Typically, parietal functions such as visuospatial processing and arithmetic calculations are unaffected even late in the FTD syndromes. Many patients with nonfluent aphasia or bvFTD later develop aspects of PSP-RS as disease spreads into subcortical or brainstem structures, or CBS-like features appear as disease moves into perirolandic cortices.

GENETIC CONSIDERATIONS