Rare forms of dementia:
GLOSSARY

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Acalculia
Inability to perform mathematical computations.

Acetylcholine
One of a group of chemicals known as neuro-transmitters. Found throughout the brain, acetylcholine enables nerve cells to communicate with each other. In Alzheimer’s disease, the levels of acetylcholine are lower than usual.

Activities of daily living
Activities necessary for everyday living, such as eating, bathing, grooming, dressing and toileting. Also referred to as basic activities of daily living (BADLS). Differs from instrumental activities of daily living (IADLS).

AD
See Alzheimer's disease

ADL
See activities of daily living

ADLs
See activities of daily living

Aetiology
The cause or origin of a disease or disorder.

Agonist
A chemical or drug that mimics the action of a neurotransmitter by stimulating the target site or receptor

Alzheimer's disease
A progressive, neurodegenerative disease characterised by loss of function and death of nerve cells in several areas of the brain, leading to loss of mental functions such as memory and learning. Alzheimer’s disease is the most common cause of dementia.

Amyloid
A protein that is found in the brains of people with Alzheimer's disease. It is deposited throughout the brain in microscopic clumps known as plaques. Its function is unknown and it may be the cause of the deterioration of brain function.

Amyloid plaque
Abnormal cluster of dead and dying nerve cells, other brain cells, and amyloid protein fragments. Amyloid plaques are one of the characteristic structural abnormalities found in the brains of individuals with Alzheimer's. Upon autopsy, the presence of amyloid plaques and neurofibrillary tangles is used to positively diagnose Alzheimer’s.

Amyloid precursor protein
A protein found in the brain, heart, kidneys, lungs, spleen, and intestines. The normal function of APP in the body is unknown. In Alzheimer's disease, APP is
abnormally processed and converted to beta amyloid protein. Beta amyloid is the protein deposited in amyloid plaques.

**antibodies**
Specialised proteins produced by the cells of the immune system that counteract a specific foreign substance. The production of antibodies is the first line of defense in the body’s immune response.

**anticholinergic drugs**
A term for drugs that reverse or inhibit the action of acetylcholine on nerve cells.

**anticholinesterase**
A class of drugs frequently prescribed to patients with Alzheimer’s disease.

**anticholinesterase drugs**
Also known as cholinesterase inhibitors, these dementia drugs, stop the breakdown of acetylcholine. They may help to slow down the progression of Alzheimer’s disease in some people. Aricept and Exelon are examples.

**anti-inflammatory drugs**
Drugs that reduce inflammation by modifying the body’s immune response.

**anxiety**
A feeling of apprehension, fear, nervousness, or dread accompanied by restlessness or tension.

**apathy**
Lack of interest, concern, or emotion.

**aphagia**
Not able to swallow.

**aphasia**
Difficulty understanding the speech of others and/or expressing oneself verbally.

**apolipoprotein E**
A protein whose main function is to transport cholesterol. The gene for this protein is on chromosome 19 and is referred to as APOE. There are three forms of APOE: e2, e3, and e4. APOE-e4 is associated with about 60 percent of late-onset Alzheimer’s cases and is considered a risk factor for the disease.

**APP**
See amyloid precursor protein.

**Aricept**
An dementia drug whose generic name is donepezil.

**atrophy**
Shrinking of size; often used to describe the loss of brain mass seen in Alzheimer’s disease during autopsy.

**autoimmunity**
Autoimmunity is when the body’s natural defences (the immune system) mistakenly attacks the body’s own tissue. "Auto" is derived from the Greek auto, meaning self, and autoimmune means attacking self.

**autonomy**
A person’s ability to make independent choices.

**autopsy**
Examination of a body organ and tissue after death. Autopsy is often performed (upon request) to confirm a diagnosis of Alzheimer’s disease.

**autosomal**
Pertaining to a chromosome not involved in sex determination. The gender does not influence the chance of inheriting the disease.

**autosome**
A chromosome not involved in sex determination. The diploid human genome consists of 46 chromosomes, 22 pairs of autosomes, and 1 pair of sex chromosomes (the X and Y chromosomes).

**axon**
The arm of a nerve cell that normally transmits outgoing signals from one cell body to another. Each nerve cell has one axon.

**B**

**Babinski’s signs**
When the sole of the foot is scratched, the big toe goes up instead of down.

**behavioural symptoms**
In Alzheimer’s disease, symptoms that relate to action or emotion, such as wandering, depression, anxiety, hostility and sleep disturbances.

**beta amyloid**
A specific type of amyloid normally found in humans and animals. In Alzheimer’s disease, beta amyloid is abnormally processed by nerve cells and becomes deposited in amyloid plaques in the brains of persons with the disease.

**biomarker**
Used to indicate or measure a biological process (for instance, levels of a specific protein in blood or spinal fluid, genetic mutations, or brain abnormalities observed in a PET scan or other imaging test). Detecting biomarkers specific to a disease can aid in the identification, diagnosis, and treatment of affected individuals and people who may be at risk but do not yet exhibit symptoms.

**brain**
One of the two components of the central nervous system, the brain is the center of thought and emotion. It is responsible for the coordination and control of bodily activities, and the interpretation of information from the senses (sight, hearing, smell, etc.). The other component of the central nervous system is the spinal cord.

**brain scan**
A general term to mean any investigation that produces pictures of the brain. A CT scan or MRI scan shows slices through the brain. A SPECT scan shows the brain’s blood supply.

**Bradykinesia**
Slowness of all voluntary movement and speech.
**BSE**  
Bovine Spongiform Encephalopathy

**bulbar**  
The bulbar region of the brain is the brainstem, the nerves coming out of the brainstem are the bulbar nerves and the muscles they innervate are the bulbar musculature. Bulbar functions include eye movements, muscle of facial expression, speaking, and swallowing.

**C**

**caregiver**  
The primary person in charge of caring for an individual having dementia, usually a family member or a designated health care professional.

**cataract**  
The development of cloudiness of the human lens due to discoloration of cells.

**cell**  
The fundamental unit of all organisms; the smallest structural unit capable of independent functioning. In the brain and nervous system important cells are the neuronal cells, which make up the nerves and brain.

**cell body**  
In nerve cells, the central portion from which axons and dendrites sprout. The cell body controls the life-sustaining functions of a nerve cell.

**cell membrane**  
The outer boundary of the cell. The cell membrane helps control what substances enter or exit the cell.

**central nervous system (CNS)**  
The part of the human nervous system consisting of the brain and spinal cord. The CNS is the control network for the entire body.

**cerebellum**  
The portion of the brain in the back of the head between the cerebrum and the brain stem. The cerebellum controls balance for walking and standing, and other complex motor functions.

**cerebral cortex**  
The outer layer of the brain, consisting of nerve cells and the pathways that connect them. The cerebral cortex is the part of the brain in which thought processes take place. In Alzheimer's disease, nerve cells in the cerebral cortex degenerate and die.

**cerebrospinal fluid (CSF)**  
The fluid that fills the areas surrounding the brain and spinal cord. It contains substances that when analyzed can help in the diagnosis of Alzheimer's disease. Collected by lumbar puncture.

**choline**  
A natural substance required by the body that is obtained from various foods, such as eggs; an essential component of acetylcholine.
choline acetyltransferase (CAT)
An enzyme that controls the production of acetylcholine; appears to be depleted in the brains of individuals with Alzheimer's disease.

cholinergic system
The system of nerve cells that uses acetylcholine as its neurotransmitter and is damaged in the brains of individuals with Alzheimer's.

cholinesterase
An enzyme that breaks down acetylcholine, into active parts that can be recycled.

chromosome
The structures within cells made up of DNA. Each chromosome carries many individual genes. Normally, human cells contain 22 pairs of chromosomes and one X and one Y or two X chromosomes depending on gender.

clinical trials
Human experiments conducted by researchers that test the value, safety and efficiency of various treatments, such as drugs.

clonus
Involuntary movement of rapidly alternating contraction and relaxation of a muscle. Ankle Clonus is the most common form of Clonus.

coexisting illness
A medical condition that exists simultaneously with another, such as arthritis and dementia.

Cognex
An anticholinesterase drug whose generic name is tacrine.

cognition
Brain functions involving thinking, remembering, learning, reasoning and planning.

cognitive abilities
Mental abilities such as judgment, memory, learning, comprehension and reasoning.

cognitive symptoms
Dysfunction of cognition, in Alzheimer's disease patients these are the defining early symptoms such as loss of memory, confusion and aphasias.

computed tomography scan (CT scan)
(pronounced "cat scan") - A type of imaging scan (X-ray) that shows the internal structure of a person's brain. In diagnosing dementia, CT scans can reveal tumors and small strokes in the brain.

D

deficits
Physical and/or cognitive skills or abilities that a person has lost, has difficulty with, or can no longer perform due to his or her dementia.
delirium
A temporary condition with rapid onset consisting of cognitive dysfunction, different from dementia in its time course.

dementia
The loss of intellectual functions (such as thinking, remembering and reasoning) of sufficient severity to interfere with a person's daily functioning. Dementia is not a disease itself but rather a group of symptoms that may accompany certain diseases or conditions. Symptoms may also include changes in personality, mood and behaviour. Dementia is irreversible when caused by disease or injury but may be reversible when caused by drugs, alcohol, hormone or vitamin imbalances or depression.

dendrites
Branched extensions of the nerve cell body that receive signals from other nerve cells. Each nerve cell usually has many dendrites.

deoxyribonucleic acid
The basis of all genetic material. Nucleotides are the building blocks of deoxyribonucleic acid (DNA). Specific patterns of nucleotides represent particular genes.

diagnosis
The process by which a physician determines what disease a patient has by studying the patient's symptoms and medical history and analysing any tests performed (blood, urine, brain scans, etc.).

disinhibition
Loss of the feelings of shame or embarrassment that normally help control a person's actions. Disinhibition results in inappropriate or improper behaviour.

disorientation
A state in which someone loses their awareness of time and place. For example, they may fail to recall the date or even the year, and may not be able to say where they are.

DNA
The basis of all genetic material. Nucleotides are the building blocks of DNA. Specific patterns of nucleotides represent particular genes.

dominant
Dominant gene (or dominant allele) is a gene which, when present, produces a certain trait, and "dominates" over a recessive allele in the gene pair.

Donepezil
An anticholinesterase drug whose generic name is Aricept.

dopaminergic
Relating to nerve cells or fibres that employ dopamine as their neurotransmitter.

dysarthria
a neurologic speech disorder caused by paralysis, weakness, improper muscle tone or incoordination of the muscles of the mouth. Dysarthria is not a disorder of language.

dysphagia
Difficulty in swallowing.
dysphasia
Lack of coordination in speech, and failure to arrange words in an understandable way; due to brain lesion. Aphasia is the complete or near complete absence of speech, and is used to describe a more severe situation than dysphasia.

E

early-onset Alzheimer’s disease
An unusual form of Alzheimer’s in which individuals are diagnosed with Alzheimer’s before the age of 65. Less than 10 percent of all Alzheimer patients have early-onset. Early-onset Alzheimer’s is associated with mutations in genes located on chromosomes 1, 14 and 21.

early stage
The beginning stages of dementia when an individual experiences very mild to moderate cognitive impairments.

enzyme
A protein produced by living organisms that promotes or otherwise influences chemical reactions.

estrogen
A hormone produced by the ovaries and testes. It stimulates the development of secondary sexual characteristics and induces menstruation in women. Estrogen is important for the maintenance of normal brain function and development of nerve cells. Estrogen is used therapeutically to treat breast and prostate cancer, osteoporosis and to relieve the discomforts of menopause. Some research suggests that estrogen may be beneficial in preventing Alzheimer’s disease. More studies are needed to confirm this.

excitotoxic
Exciting neurons which can over time lead to neuronal death.

executive function
The ability to plan actions and change plans when adaptation is necessary

F

familial Alzheimer’s disease (FAD)
A form of Alzheimer’s disease that runs in families.

fasciculation
A small local contraction of muscles, visible through the skin

free radicals
Highly reactive molecules capable of causing damage in brain and other tissue. Free radicals are common by-products of normal chemical reactions occurring in cells. The body has several mechanisms to deactivate free radicals.
gait
A person’s manner of walking. People in the later stages of Alzheimer’s often have "reduced gait," meaning they may lose the ability to lift their feet as they walk.

gene
The basic unit of heredity; a section of DNA coding for a particular trait.

gene linkage
A group of genes located closely together on a chromosome. Used by researchers to related diseases to specific genes.

gene regulation
The control of the rate or manner in which a gene is expressed as a protein.

 genetic susceptibility
The state of being more likely than the average person to develop a disease as a result of genetics.

 genome
All the genes of an organism. The Human Genome Project is currently trying to map all of the genes of the human genome by the year 2003.

glial
Pertaining to the supporting cells of neural tissue.

 glial cells
Glial cells are maintenance and support cells in the central nervous system (CNS). There are a number of different types of glial cell in the CNS including: oligodendrocytes, astrocytes and microglia.

 gliosis
In the brain, scars are formed by glial cells and are called glial scars or gliosis.

 glucose
A simple sugar that is a major energy source for all cellular and bodily functions. Glucose is obtained through the breakdown or metabolism of food in the digestive system.

 glutamate
An amino acid neurotransmitter normally involved in learning and memory. Under certain circumstances it can be an excitotoxin and appears to cause nerve cell death in a variety of neurodegenerative disorders.

hallucination
A sensory experience in which a person can see, hear, smell, taste or feel something that isn’t there.

 heavy metals
The term heavy metal refers to any metallic chemical element that has a relatively high density and is toxic, highly toxic or poisonous at low concentrations. Examples of heavy metals include mercury (Hg), cadmium (Cd), arsenic (As), chromium (Cr), thallium (Tl), and lead (Pb).

**hippocampus**
A part of the brain that is important for learning and memory.

**hypokinesia**
Decreased muscular activity.

**immune system**
A system of cells that protect a person from bacteria, viruses, toxins and other foreign substances that enter the body.

**incontinence**
Loss of bladder and/or bowel control.

**inflammatory response**
The immune system’s normal response to tissue injury or abnormal stimulation caused by a physical, chemical or biological substance. Immune system cells, if abnormally stimulated, can often cause further tissue damage while responding to the injured site.

**lability**
Unstable, easily changed. The word is usually applied to rapid mood swings.

**lack of coordination**
Uncoordinated movement is an abnormality of muscle control or an inability to finely coordinate movements, resulting in a jerky "to-and-fro" unsteady motion of the trunk or the limbs.

**late-onset Alzheimer’s disease**
The most common form of Alzheimer’s disease, usually occurring after age 65. Late-onset Alzheimer’s strikes almost half of all people over the age of 85 and may or may not be hereditary.

**late stage**
Designation given when dementia symptoms have progressed to the extent that a person has little capacity for self-care.

**lumbar puncture**
A procedure used to collect cerebrospinal fluid, which can help in the diagnosis of Alzheimer’s disease, also called spinal tap.
magnetic resonance imaging
A brain scanning technique that generates cross-sectional images of a human brain by detecting small molecular changes. MRI scans reveal a contrast between normal and abnormal tissues. The image produced is similar to those generated by CT scans. There are no side effects or risks associated with MRI scans, although MRI can affect electrical devices like pacemakers and hearing aids.

mania
A state characterised by a pervasive and abnormally expansive mood, elation, irritability, flight of ideas, pressured speech and increased motor activity.

memory
The ability to process information that requires attention, storage and retrieval.

metabolism
The complex chemical and physical processes of living organisms that promote growth, sustain life and enable all other bodily functions to take place.

Mini-Mental State Examination
A standard mental status exam routinely used to measure a person’s basic cognitive skills, such as short-term memory, long-term memory, orientation, writing, and language.

mitochondria
Components found in cells that serve as primary energy sources for all cellular functions.

model system
A system used to study processes that take place in humans or other living organisms.

monogenic
Controlled by or associated with a single gene.

muscular dystrophies
A group of genetic degenerative myopathies characterized by weakness and muscle atrophy without nervous system involvement.

mutation
A sudden change in the genetic constitution.

myoclonus
A brief, shock like contraction of a single muscle or of one or more muscle groups, rarely of a part of a muscle.

myopathy
Disease of the muscle tissues, which include the muscles over our bones (skeletal muscle) and the heart (cardiac muscle).

nerve cell (neuron)
The basic working unit of the nervous system. The nerve cell is typically composed of a cell body containing the nucleus, several short branches (dendrites), and one long arm (the axon) with short branches along its length and
at its end. Nerve cells send signals that control the actions of other cells in the body, such as other nerve cells and muscle cells.

**neuro-degenerative disease**
A type of neurological disorder marked by the loss of nerve cells.

**neurofibrillary tangle**
Accumulation of twisted protein fragments inside nerve cells. Neurofibrillary tangles are one of the characteristic structural abnormalities found in the brains of Alzheimer patients. Upon autopsy, the presence of amyloid plaques and neurofibrillary tangles is used to positively diagnose Alzheimer’s.

**neurological disorder**
Disturbance in structure or function of the nervous system resulting from developmental abnormality, disease, injury, or toxin.

**neuron**
See nerve cell.

**neuropathology**
Changes in the brain produced by a disease.

**neurotransmission**
Passage of signals from one nerve cell to another via chemical substances or electrical signals.

**neurotransmitter**
Specialised chemical messenger (e.g., acetylcholine, dopamine) that sends a message from one nerve cell to another. Most neurotransmitters play different roles throughout the body, many of which are not yet known.

**nucleotides**
The different building blocks of DNA, represented by the letter A, T, G and C.

**nucleus**
The central component of a cell. It contains all genetic material.

**O**

**onset**
Defines time of life when disease begins (e.g., early-onset, late-onset).

**overvalued ideas**
Unreasonable and persistent beliefs, held with less than delusional intensity, which are not generally held in the patient's culture. Overvalued ideas may have a basis in reality, such as preoccupations that one's nose is too large, that only diet can cure cancer, or that "having a baby is the only way I'll ever be happy." Ideas of reference are one type of overvalued idea.

**P**

**paranoia**
Suspicion of others that is not based on fact.
paranoid idea
An overvalued idea that one is being persecuted.

paraparesis
Weakness affecting the lower extremities (the hip, thigh, leg, ankle, and foot)

parkinsonism
A group of neurological disorders characterised by hypokinesia, tremor and muscular rigidity.

Parkinson’s disease
A disorder of the brain characterised by shaking (tremor) and difficulty with walking, movement, and coordination. The disease is associated with damage to a part of the brain that is involved with movement.

penetrance
An individual who carries a dominant gene may show a variable degree of the symptoms of the disorder.

PET scan
See positron emission tomography scan.

phenotype
Expression of any of those genes as a physical, biochemical or physiological trait.

plaques
See amyloid plaque.

positron emission tomography scan (PET scan)
An imaging scan that measures the activity or functional level of the brain by measuring its use of glucose.

presenilins
Proteins that may be linked to early-onset Alzheimer’s disease. Genes that code for presenilin 1 and presenilin 2 have been found on chromosomes 14 and 1, respectively, and are linked to early-onset familial Alzheimer’s disease.

prions
Protein segments that may cause infection that may lead to some forms of dementia.

Protein
The product of gene expression. Proteins are the molecules that do much of the work in the body such as creating structures, utilising and storing energy and transmitting signals.

psychosis
A general term for a state of mind in which thinking becomes irrational and/or disturbed. It refers primarily to delusions, hallucinations, and other severe thought disturbances.

recessive
Recessive gene (or recessive allele) is a gene, which must be present on both chromosomes in a pair to show outward signs of a certain characteristic.

**repetitive behaviors**
Repeated questions, stories and outbursts or specific activities done over and over again, common in people with dementia.

**riluzole**
A drug that has been shown to have energy buffering and anti-glutamate properties

**risk factors**
Factors that have been shown to increase one’s odds of developing a disease. In Alzheimer’s disease, the only established risk factors are age, family history and genetics.

**S**

**senile plaque**
See amyloid plaque.

**side effect**
An undesired effect of a drug treatment that may range in severity from barely noticeable, to uncomfortable, to dangerous. Side effects are usually predictable.

**SPECT scan**
A painless procedure that takes a picture of a person's brain, which can help in the diagnosis of Alzheimer’s disease, yields somewhat different information from MRI.

**Spinal cord**
One of the two components of the central nervous system, the spinal cord carries signals between the brain and the rest of the body to allow a person to sense the environment and react to it. The other component of the central nervous system is the brain.

**sporadic**
Occurring occasionally in a random or isolated manner.

**stages**
Course of disease progression defined by levels or periods of severity: early, mild, moderate, moderately severe, severe.

**synapse**
The junction where a signal is transmitted from one nerve cell to another, usually by a neurotransmitter.

**T**

**Tacrine**
An anticholinesterase drug, also called Cognex.
tangles
See neurofibrillary tangles.

tauopathies
Mutations on tau gene are directly causing a disease named “fronto-temporal dementia with parkinsonism linked to chromosome 17” (FTDP-17). These mutations (more than 30) have demonstrated the important role of tau pathology in neurodegenerative disorders. Indeed, more than 20 neurodegenerative disorders have a tau pathology, generally with an accumulation of tau proteins in neurons or glial cells, or both. The fact that tau can be directly responsible of diseases and that most dementing disorders have a tau pathology has generated this concept. Tauopathies comprise primary and secondary tauopathies. Primary tauopathies are the diseases with tau playing a major role, such as FTDP-17, PSP, CBD and most fronto-temporal dementia. Secondary tauopathies are diseases like Alzheimer's disease, which is likely a true tauopathy, but fuelled by defects of the APP metabolic pathway.

tau protein
The major protein that makes up neurofibrillary tangles found in degenerating nerve cells. Tau is normally involved in maintaining the internal structure of the nerve cell. In Alzheimer’s disease, tau protein is abnormally processed.

tissue
A group of similar cells that act together in the performance of a particular function.

toxin
A substance that can cause illness, injury or death. Toxins are produced by living organisms.

trigger
An environmental or personal stimulus that sets off particular and sometimes challenging behavior.

U

upper motor neuron signs
Signs and symptoms that result from damage to descending motor systems. These include paralysis, spasticity and a positive Babinski sign (reflex).
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