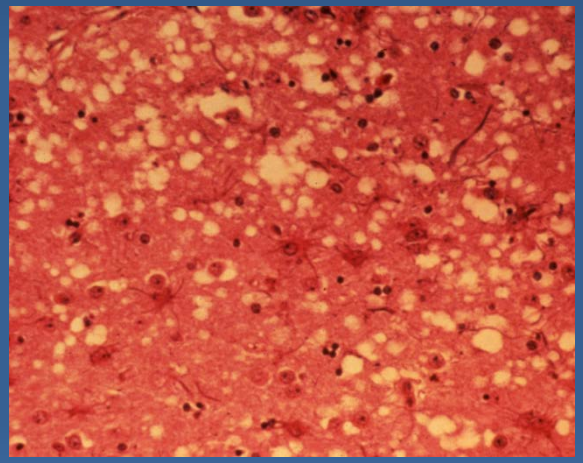

Palliative management of 14 patients with Creutzfeldt – Jakob Disease (CJD)

Ms Danielle Bach
Ms Jo-Anne Wilkinson
Dr Sonia Fullerton

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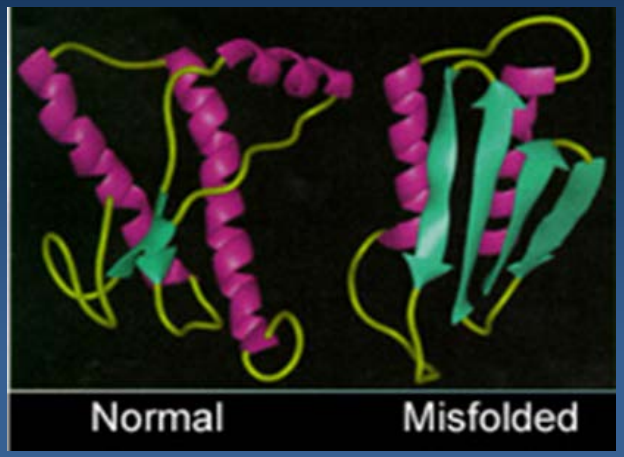
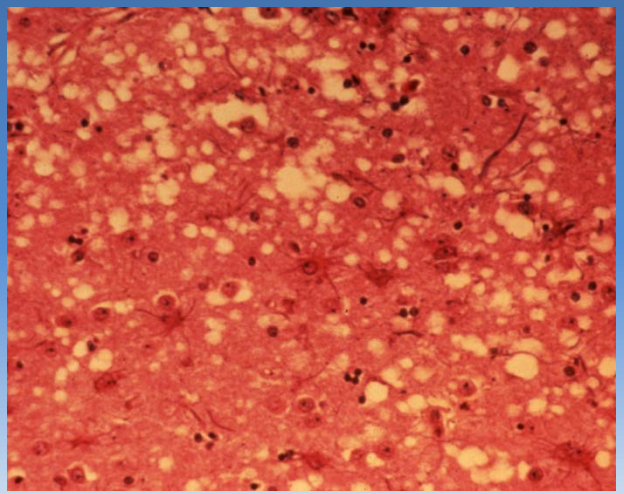
CJD:
"Proteins behaving badly"



- 'TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHY'
- RARE - one in a million, 35 cases per yr in Aus
- FATAL
- RAPID DEGENERATION – "Dementia on fast forward"
- AUTOPSY for confirmation
- TENACIOUS
- HUMANS & ANIMALS



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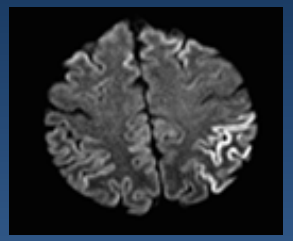
Types of CJD



- Sporadic - (85 - 90%) just happens!
  - 50 - 70 year age group, survival 3 - 6 mths
- Genetic - (10 - 15%), autosomal dominant
  - 30 - 60 year age group, survival 2 - 5 yrs
- Acquired - (1%)
  - Iatrogenic - explanted materials, surgical instruments
  - Variant - BSE

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Got it! Mad Cow Disease, right?

"CJD not tested for, very unlikely in setting of being a vegetarian"

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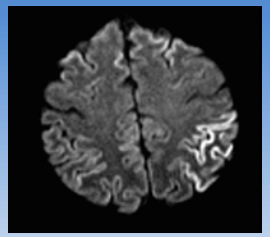
Why were we interested to look at patients with a diagnosis of presumed CJD and their families?

- 14 patients from 2011-2014
- Limited published information
- Health services – room to improve
- Issues faced by patients and families



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What issues are faced by patient and families?

- Complex, rapidly escalating symptoms
- No cure
- Unfairly stigmatised
- Sporadic vs Genetic
- Media

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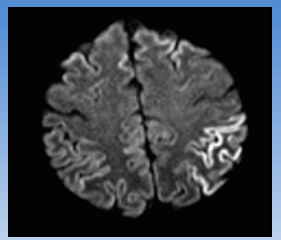


Retrospective audit of 14 patients with presumed CJD

characteristics

demographics

Diverse ethnicities & occupation

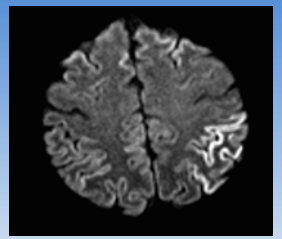
10 Male
4 Female

Nil significance

Median age 67 years
Range 54 - 58 years

comorbidities

age



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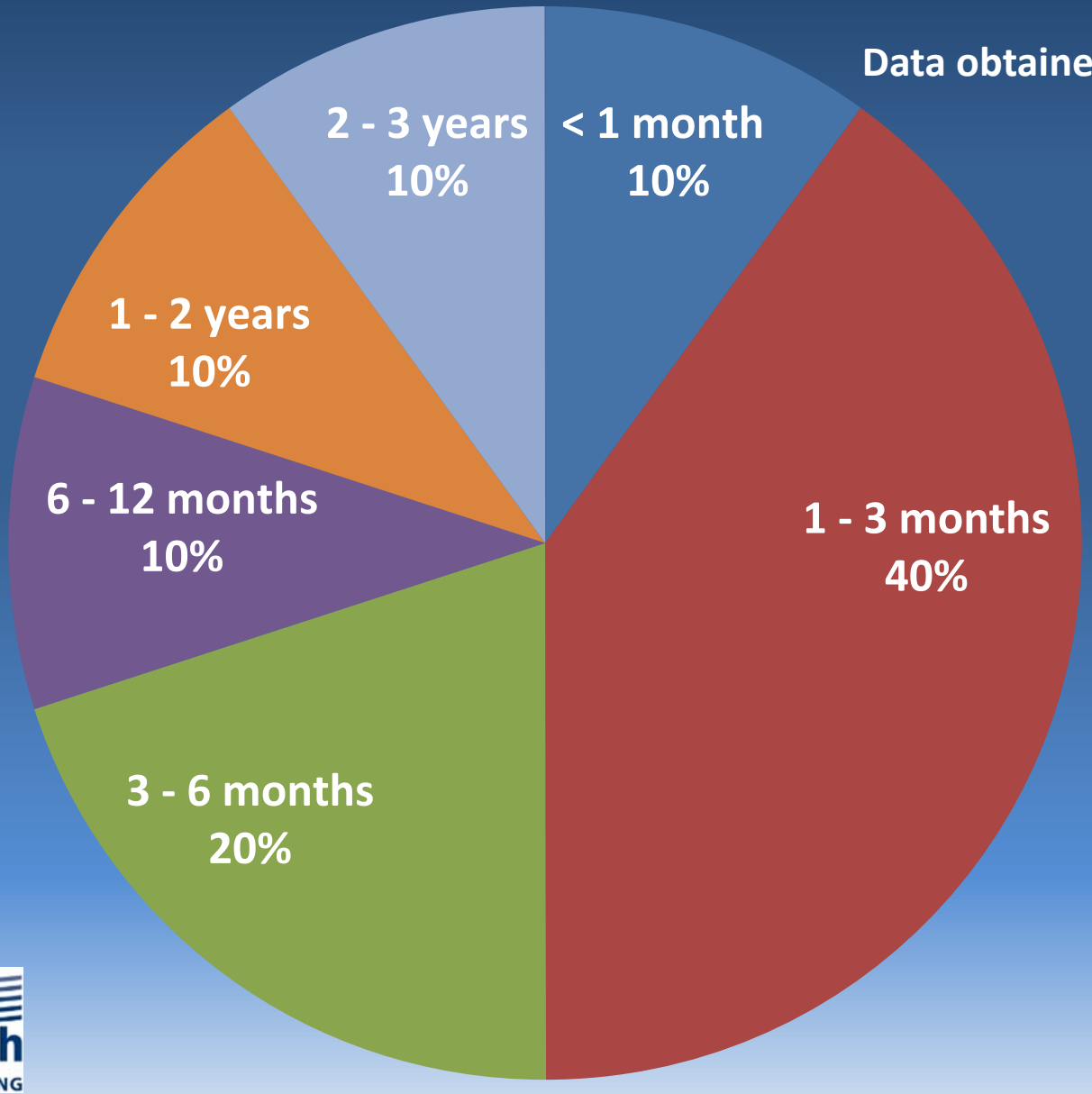
How long was the patients' journey?

Data obtained for 10 patients



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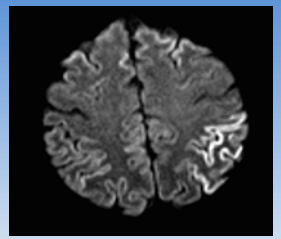

35 DAY ROLLER COASTER RIDE

- D1 PSYCH - 'bizarre behaviour', ?Psychosis, CT Brain NAD, Failure to respond to Mx, ?organic
- D15 GEN MED - 1:1, FNC, hallucinating, dysphagic, Fall, incontinent, fluctuating GCS, VCAT, Code Grey, ?Encephalitis, MRI Brain NAD, LP
- D23 NEURO - "CJD Likely"- aphasic, rigid, terror stricken, myoclonic jerks. For EEG
- D26 - Family meeting - informed likely CJD, decision for placement.

- D29 - s/b Palliative Care - rigid, non verbal, ?PCU
- D30 - Stopped eating, CTCAP & rpt MRI Brain - “Probable CJD”, active Mx cont
- D31 - Family request pall. approach, fluctuating GCS, MET Call , seizures
- D33 - s/b PCT - ‘too unwell to transfer’
- D34 - PALLIATIVE CARE UNIT - arrives late pm unresponsive
- D35 - 0500 RIP, daughter present

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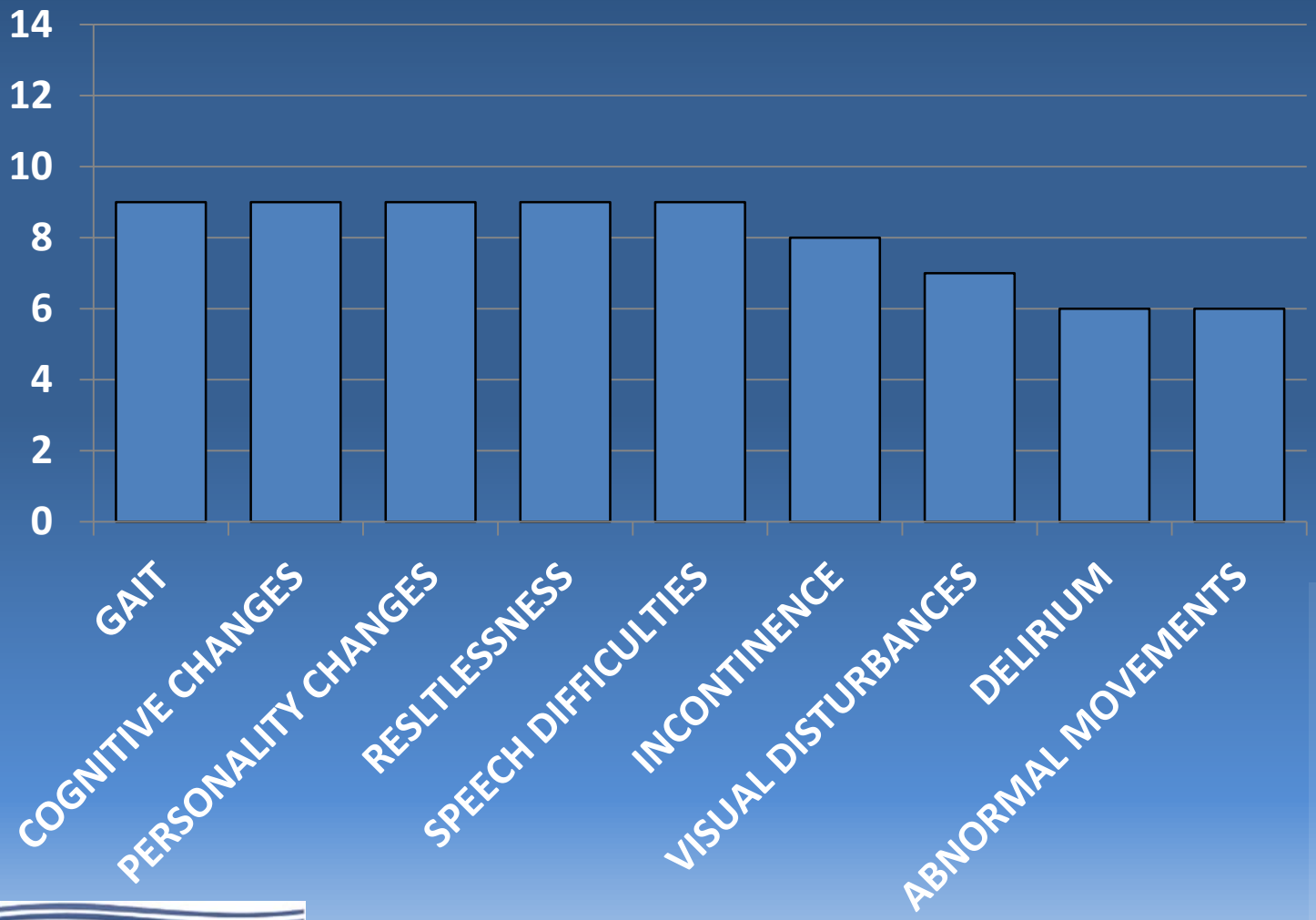
How did these patients present?



The most common symptoms –

- Gait
- Cognition
- Personality
- Restlessness
- Speech

The presenting symptoms were very diverse

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Investigations performed

INVESTIGATIONS	% OF PTS
MRI	93
LUMBAR PUNCTURE	86
CT BRAIN, SYPHILIS	57
EEG	64
HSV	43
AUTOIMMUNE, THYROID	36
HIV	36
PARANEOPLASTIC, HEAVY METAL	21
HEPATITIS, PET SCAN	14
OTHER	7

Differential Diagnoses

DIFFERENTIAL Dx	% OF PTS
DEMENTIA	50
ENCEPHALITIS, SYPHILIS	43
HSV	35
HIV	28
HEAVY METAL	28
STATUS EPILEPTICUS, PARANEOPLASTIC SYNDROME, AUTO IMMUNE	21
ACUTE BRAIN SYNDROME, HEPATITIS	14
HYDROCEPHALUS, CVA, MALIGNANCY, DRUG RELATED, FE DEF, CMV, CO2 NARCOSIS, MASTOIDITIS, WERNICKE'S SYNDROME, CEREBRAL ATAXIA	7

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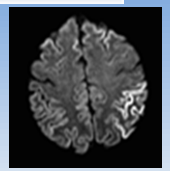
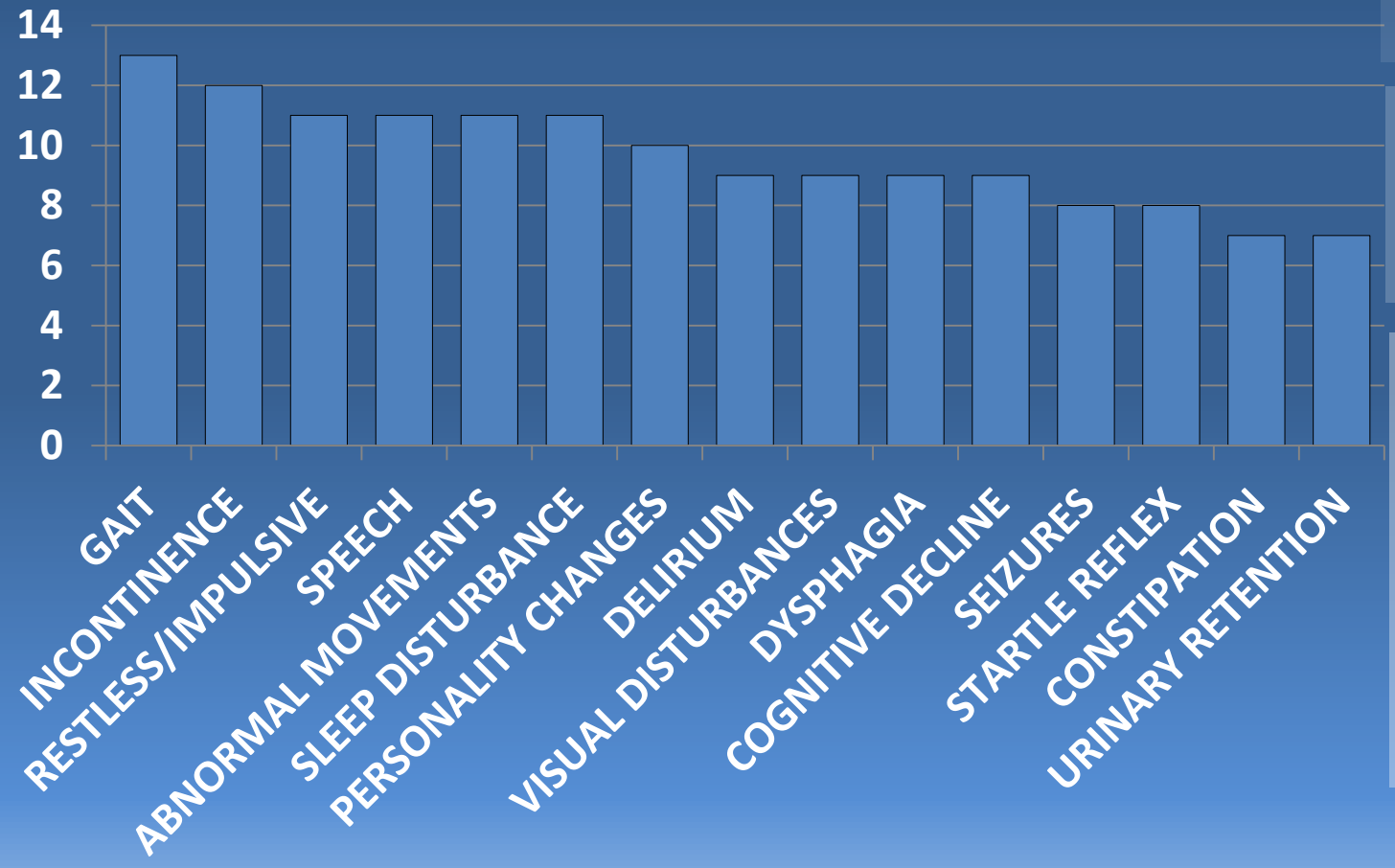


What symptoms developed as people became sicker?



93% gait disturbance

86% incontinence

80% abnormal speech, abnormal movements, sleep disturbance, personality changes

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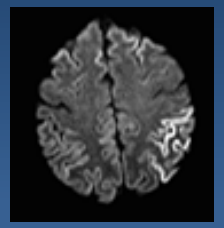
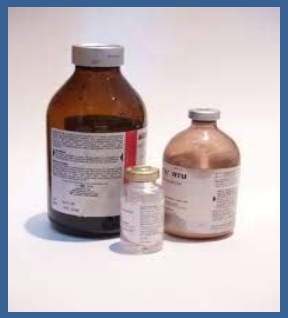
What pharmacological management did patients need?





Opioids

Anticholinergics

Benzodiazepines
Antipsychotics
Aperients

There is little information in the medical literature to guide care

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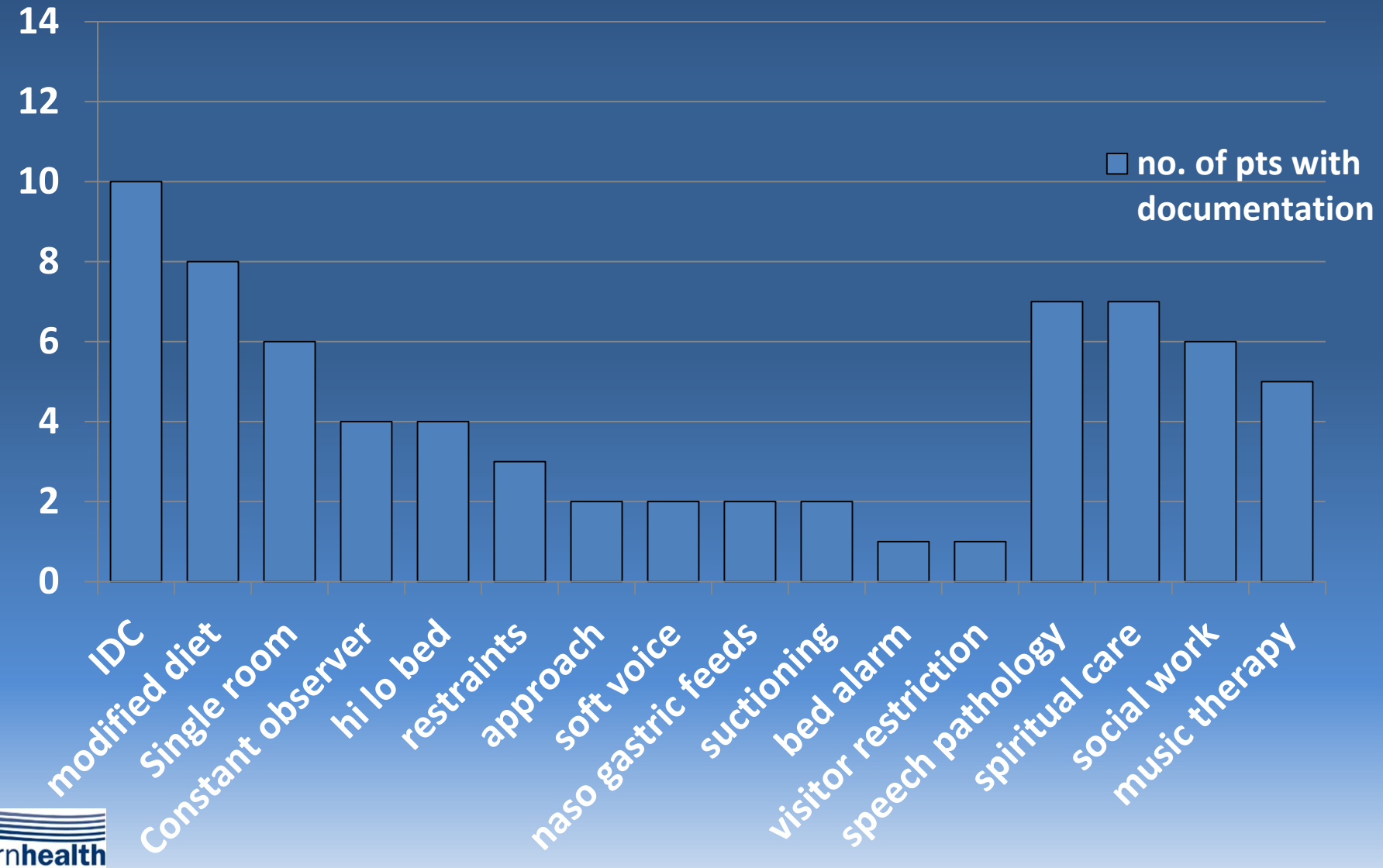
What non pharmacological interventions were documented?



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Were patients referred to the CJD support group network?

Only documented in 4 cases

Room for improvement!

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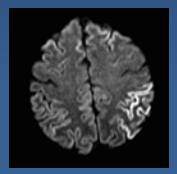
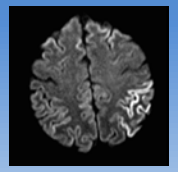
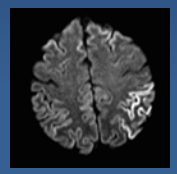
Implications for practice



There is a wide variation in symptoms experienced at presentation and throughout disease progression

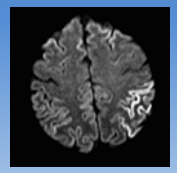
The trajectory of disease is often more rapid than we normally see in other diseases





Documentation of interventions and their effectiveness will assist timely management

Psychosocial support for the family is paramount.



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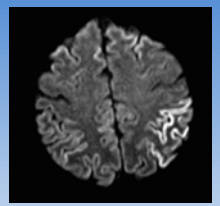

image courtesy of Dr Chris O'Donnell, Radiopaedia.org, rID: 16320