

Localisation of archetype for
Intractable disease surveillance
program in Japan

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Agenda

- Archetype localisation
 - Localisation, internationalisation, adaptation
- Localisation to Japan
 - Intractable disease surveillance program
 - Archetype localisation/adaptation
- Summary

Ehime



Localisation / Internationalisation

- Localisation
 - Language translation
 - Currency, Date, time, time zone
 - Culture, rules, law
- Internationalisation
 - Capacity for multiple localisation

Common problems in localisation to Japan

- Translation
 - Word sequence
 - Ex. From A to B(En) AからBへ(Ja)
 - Dialog box
 - Not one to one
 - By context
- Character encoding
 - UTF-8, UTF-16, S-JIS, JIS, EUC-JPN....
- Address sequence
 - Reverse from Western style
 - Prefecture, City, Town, number

Archetype in intractable disease surveillance in Japan

- Localisation
 - Archetype translation to Japanese
 - Design for Japan domestic clinical issue
- Adaptation
 - General practice <-> Specialised medical service
 - Auto-immune disease, neuron degenerative disorders
 - Archetype adaptation to Japanese clinical environment
 - Easy: Physiology, Chemistry
 - Difficult: Demographics, Administrative concept

Why openEHR?

- Interoperability for
 - Clinical research
 - Healthcare governance
 - Evaluate clinical care
- ISO 13606 Standard
 - Experience
 - 20years
 - Establishment
 - Implementation

I Love this community!

Healthcare insurance in Japan

- Universal care
 - For all patient
 - For all medical provider
- Two system
 - Medical insurance(for all generation)
 - Long term care insurance(for aged people)
- Support program
 - Handicapped, Children, Single mother...

'Nanbyo'

- So called 'intractable disease'
- Definition
 - diseases that have resulted from an unidentifiable cause and, without a clearly established treatment, have a considerably high risk of disability
 - diseases that chronically develop and require a significant amount of labor for the patient's care, causing a heavy burden on other family members of the patient, both financially and mentally

'Tokutei shikkan'

- Specified rare and intractable disease
 - Public subsidized
- Definition
 - Chronic development and serious consequences
 - Treatment of these disease is expensive for patients and families.
 - Lack of information for diseases
- Classification
 - 56 diseases, 615,568 patients registered

List of 'tokutei shikkan'

- | | | | |
|---|--|--|--|
| 01 Behcet disease | Parkinson disease | familial insomnia | abnormal secretion of prolactin - Syndrome of abnormal secretion of gonadotropin - Syndrome of abnormal secretion of antidiuretic hormone - Syndrome of abnormal secretion of Thyroid stimulating hormone - Cushing disease - Acromegaly - Hypopituitarism |
| 02 multiple sclerosis | 21 amyloidosis | 39 primary pulmonary hypertension | |
| 03 myasthenia gravis 0 | 22 ossification of posterior longitudinal ligament (OPLL) | 40 neurofibromatosis type 1, neurofibromatosis type 2 | |
| 4 systemic lupus erythematosus (SLE) | 23 Huntington disease | 41 subacute sclerosing panencephalitis (SSPE) | |
| 05 subacute myelo-optico-neuropathy (SMON) | 24 moyamoya disease | 42 Budd-Chiari syndrome | |
| 06 aplastic anemia | 25 Wegener granulomatosis | 43 idiopathic chronic pulmonary thromboembolism with pulmonary hypertension | |
| 07 sarcoidosis | 26 dilated cardiomyopathy, congestive cardiomyopathy | 44 lysosomal storage diseases - Fabry disease - other lysosomal storage diseases | 615,568 patients registered |
| 08 amyotrophic lateral sclerosis (ALS) | 27 multiple system atrophy - striatonigral degeneration (SND) - olivopontocerebellar atrophy (OPCA) -Shy-Drager syndrome | 45 adrenoleukodystrophy (ALD) | |
| 09 scleroderma , dermatomyositis, or polymyositis | 28 epidermolysis bullosa | 46 Familial Hypercholesterolemia (Homozygous type) | |
| 10 idiopathic thrombocytopenic purpura | 29 pustular psoriasis | 47 spinal muscular atrophy (SMA) | |
| 11 (1) polyarteritis nodosa (2) microscopic polyangiitis | 30 disseminated spinal canal stenosis | 48 spinal and bulbar muscular atrophy (SBMA) | |
| 12 ulcerative colitis | 31 primary biliary cirrhosis | 49 Chronic Inflammatory Demyelinating Polyneuropathy | |
| 13 Takayasu arteritis | 32 severe acute pancreatitis | 50 Hypertrophic cardiomyopathy | |
| 14 thromboangitis obliterans, Buerger disease | 33 idiopathic necrosis of the femoral head | 51 Restrictive cardiomyopathy | |
| 15 pemphigus | 34 mixed connective-tissue disease | 52 Mitochondrial disease | |
| 16 spinocerebellar degeneration | 35 primary immunodeficiency syndrome | 53 lymphangioliomyomatosis | |
| 17 Crohn disease | 36 idiopathic interstitial pneumonia | 54 Severe erythema multiforme (Acute Phase) | |
| 18 fulminant hepatitis | 37 retinitis pigmentosa | 55 Ossification of the ligamentum flavum | |
| 19 malignant rheumatoid arthritis (rheumatoid vasculitis) | 38 prion diseases -Creutzfeldt-Jakob disease (CJD) - Gerstmann-Straussler-Sheinker syndrome -fatal | 56 Diencephalo-hypophysial dysfunction -Syndrome of | |
| 20 Parkinson disease and related diseases -progressive supranuclear palsy - corticobasal degeneration - | | | |

The Specified Disease(tokutei shikkan) Treatment Research Program

- Prefecture government
 - Certificate and yearly renewal
 - Application document includes:
 - Application and consent form
 - Clinical research form certified by doctor
 - Certificate of residence, earnings, and other
- Subsidy
 - Partial-total by earnings

Problems

- Certificate criteria
 - Vary from prefecture to prefecture
- National data registry
 - Permitted researchers construct databases for their study at that time
 - Nation wide follow up system
 - Quality management
 - Epidemiological new findings
 - Security issues

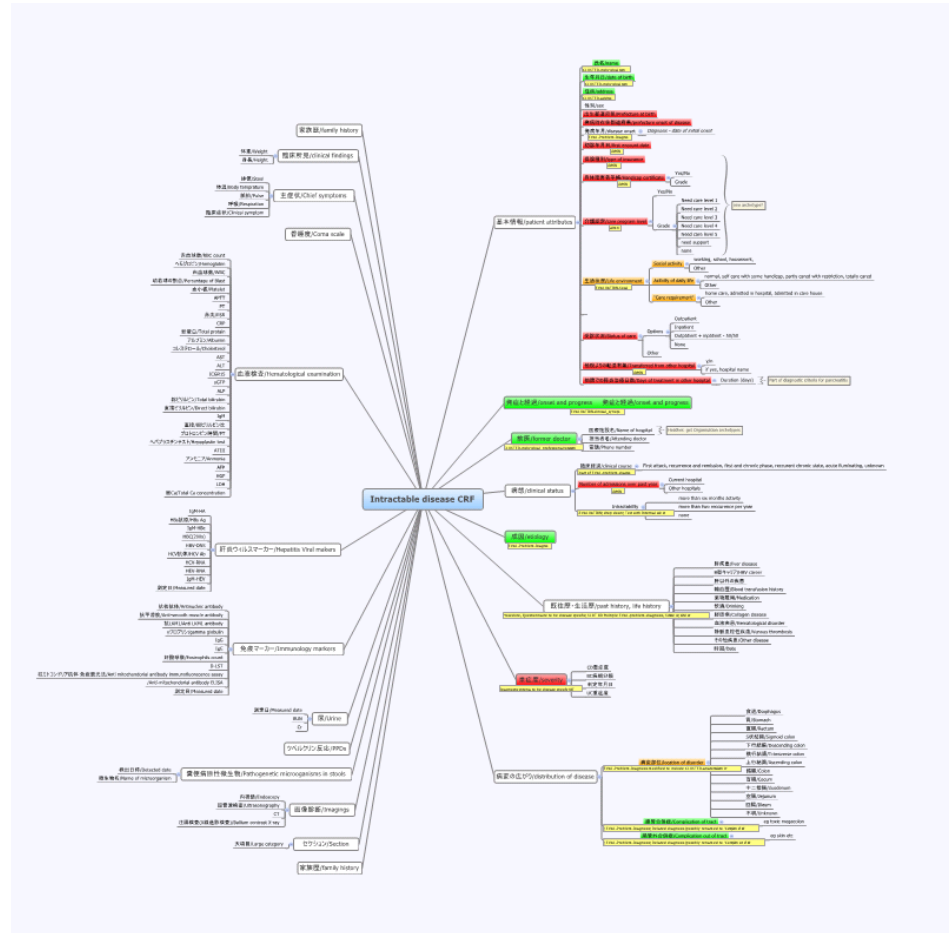
Schedule

- First year (- March 2010)
 - Modeling a few clinical research forms
 - Ulcerative colitis, Crohn disease
 - Report the problems of existing forms
- Second year(April 2010 – March 2011)
 - Implementation of server and registry
 - More diseases modeling

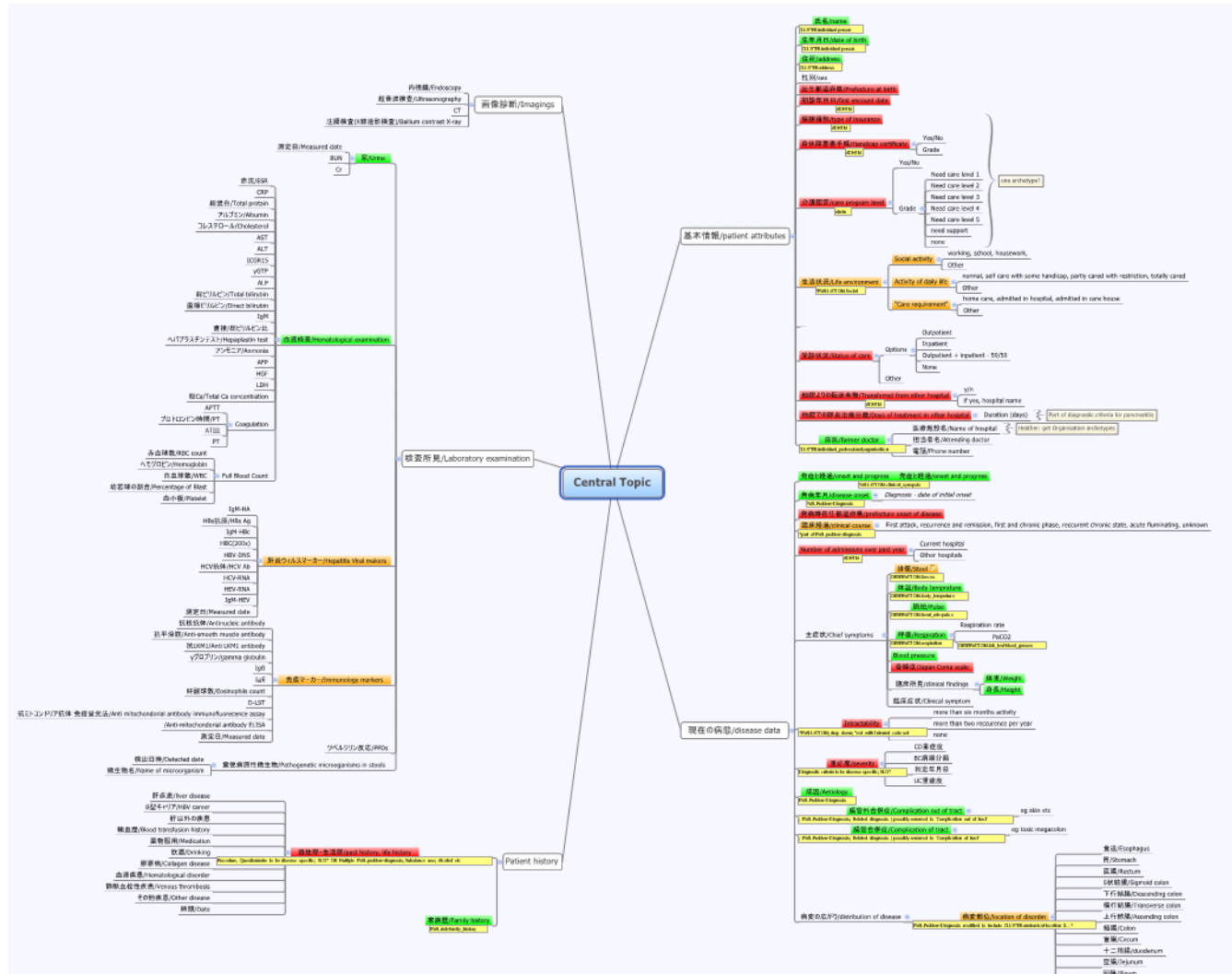
Project organization

- Leader
 - Prof. Tsutomu Chiba, Gastroenterology, Kyoto Univ
- Clinical group
 - Prof. Tsuneyo Mitsumori, Clinical immunology, Kyoto Univ
 - Prof. Ryosuke Takahashi, Clinical neurology, Kyoto Univ
- Informatics Group
 - Prof Hiroyuki Yoshihara, Kyoto Univ
 - Eizen Kimura, Shinji KOBAYASHI, Prof Ken Ishihara, Ehime Univ

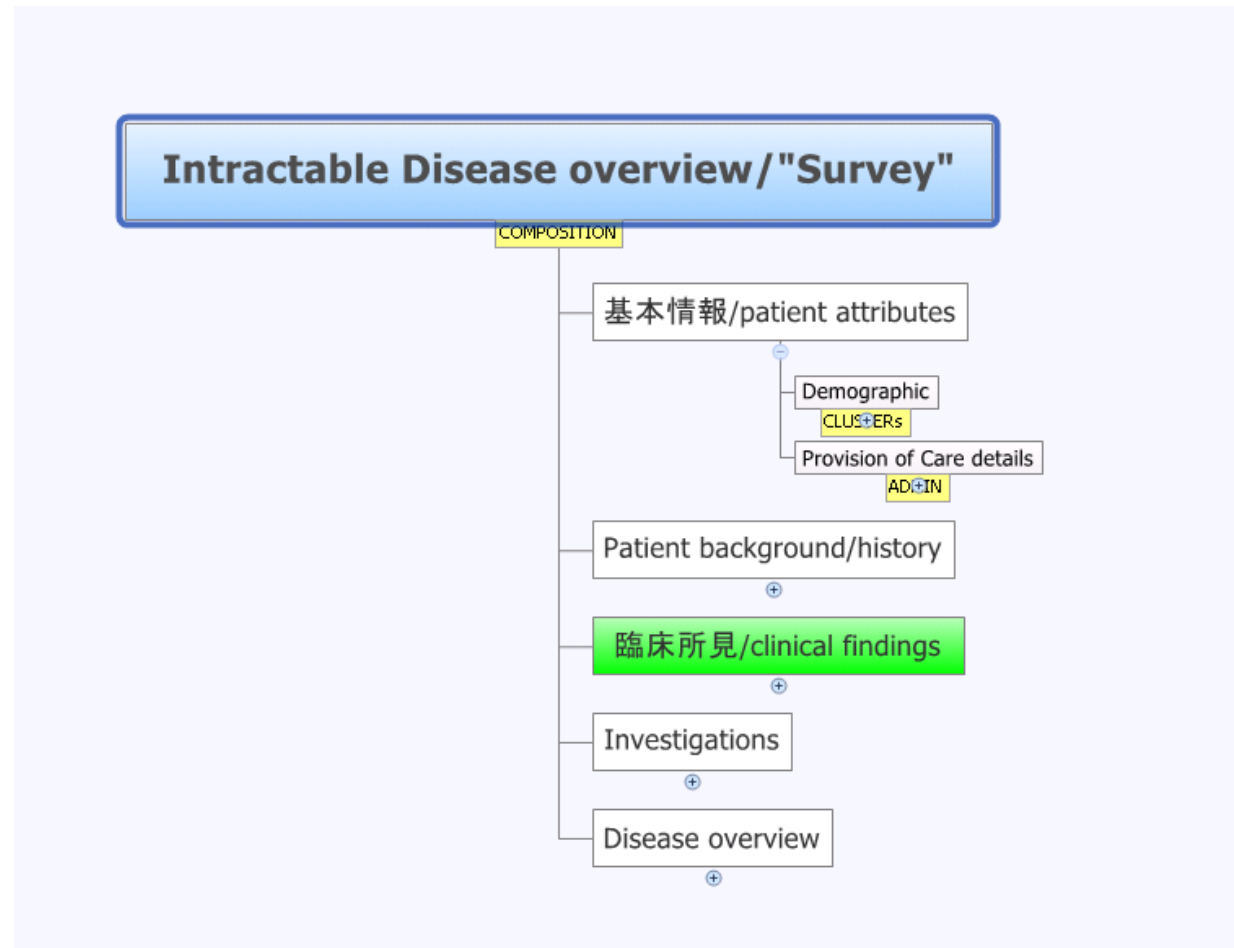
Mindmap designed for clinical research form, first trial



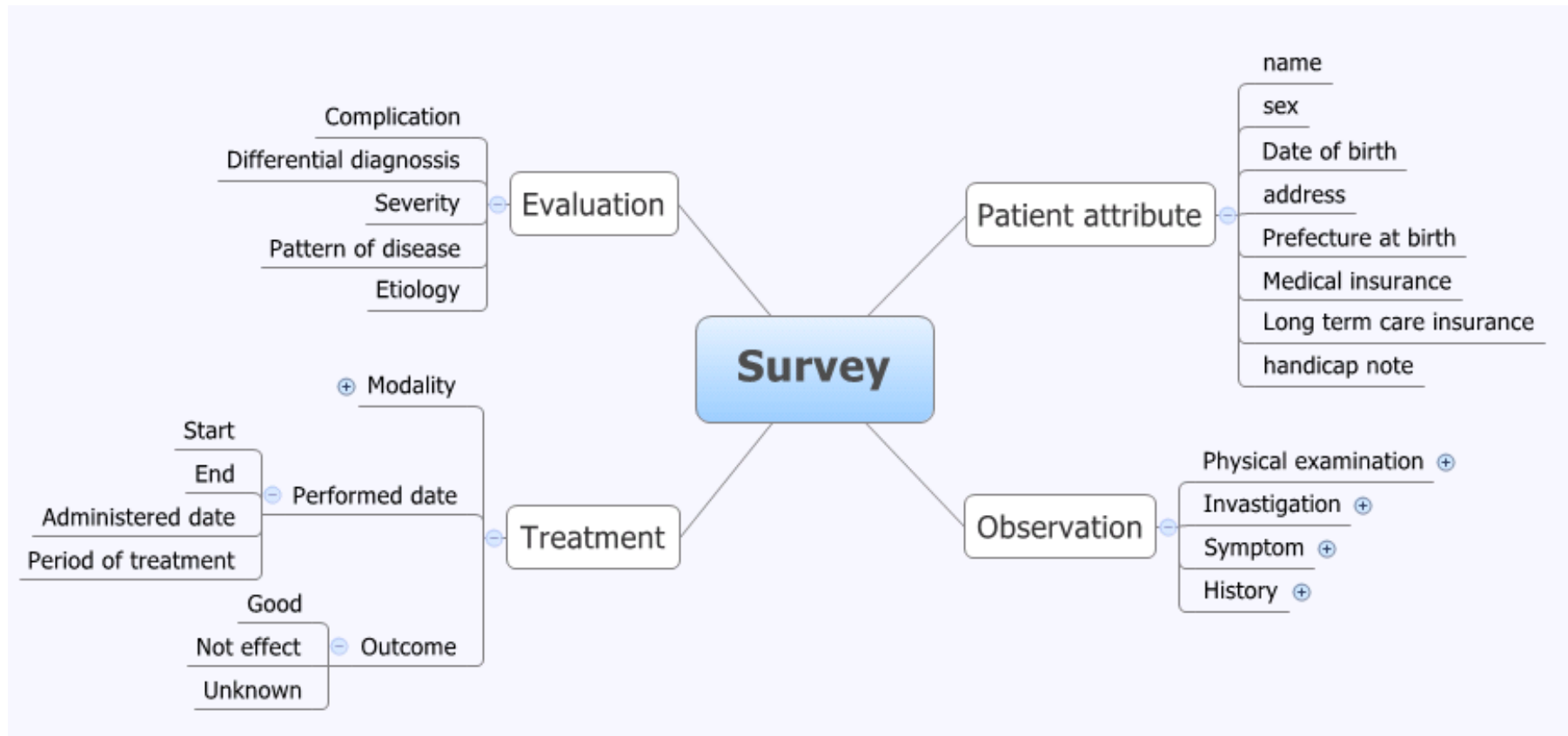
MindMap version 2



Mindmap version 3



MindMap current version



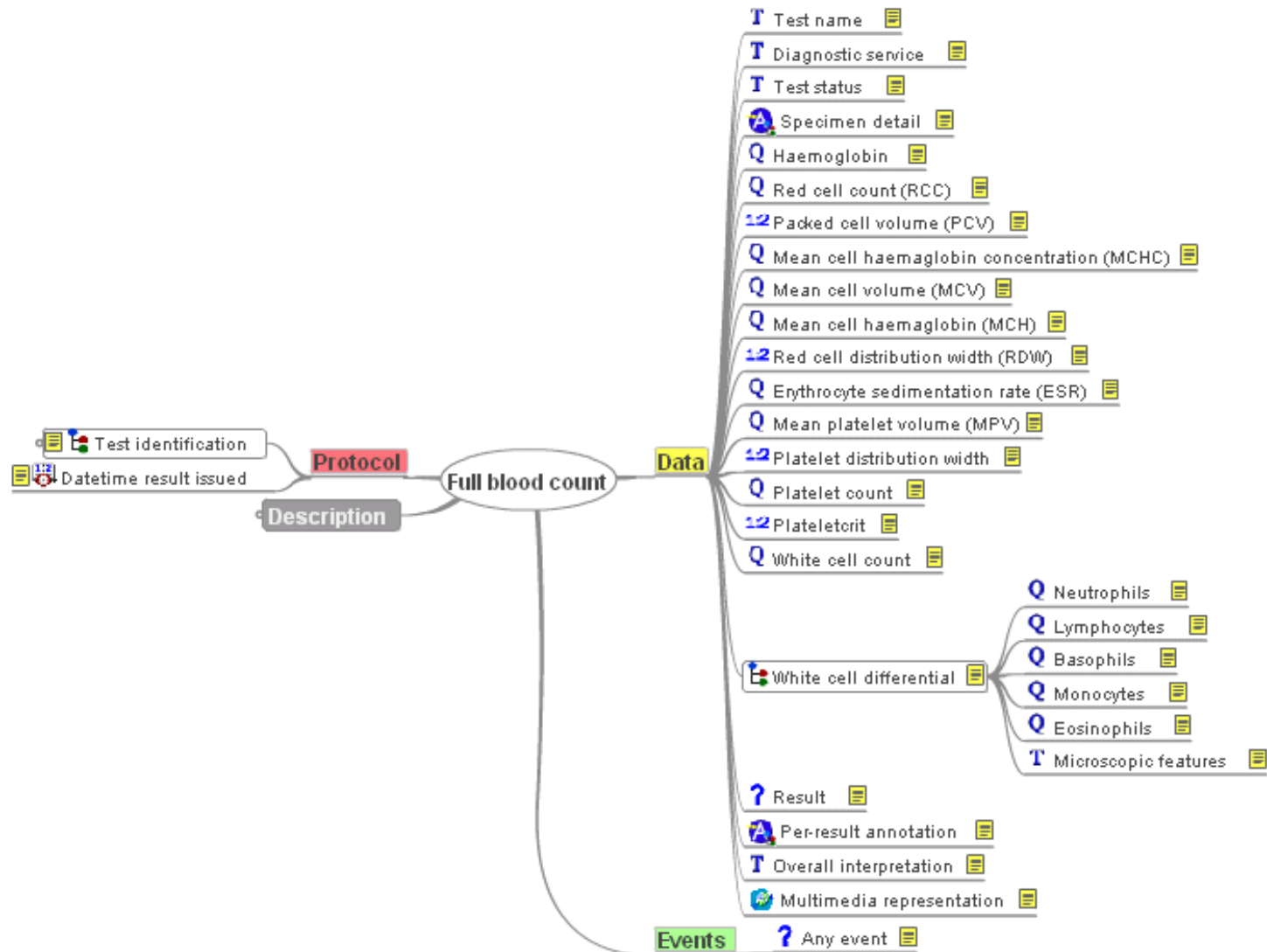
Archetype localisation/adaptation

- Address
- ESR/FBC
- Insurance
- Family history
- Severity?
- Modality?
- Terminology

Address in Nanbyo form

- Prefecture government
 - Nation budget
 - management by prefecture
 - Prefecture deal subsidy to patients
- Address archetype localisation
 - Lack of granularity for Japanese local government
 - Reversed style

FBC archetype



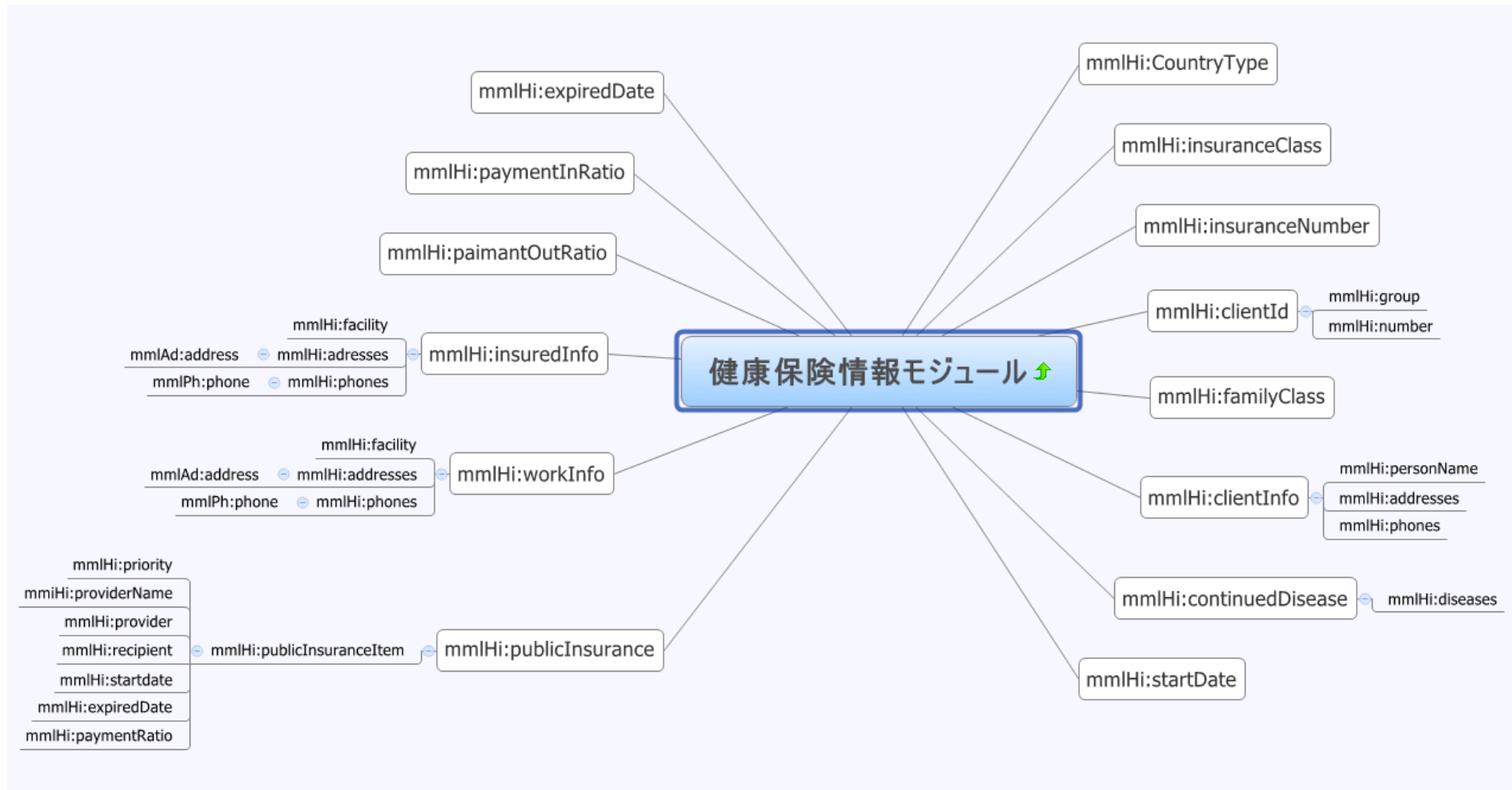
ESR/FBC

- ESR(Erythrocyte sedimentation ratio)
 - Marker for SLE activity(not CRP)
 - Included in FBC archetype
- FBC(Full blood Count)
 - WBC, RBC, Ht, Hb, Plt,(ESR)
 - WBC classification lacks immature myeloid series
- ESR is isolated from FBC
- WBC classification -> cluster?

Blood cell classification (microscopic)

- Myeloblast
- Promyelocyte
- Myelocyte
- Metamyelocyte
- Proerythroblast
- Basochromatic-
- Orthochromatic
- Normochromatic
- Megaloblast
- Erythroblast
- Reticulum
- Monoblast
- Promonocyte
- Phagocyte
- Lymphoblast
- Plasmacyte
- Megakaryocyte
- Promegakaryocyte

Health insurance(MML module)



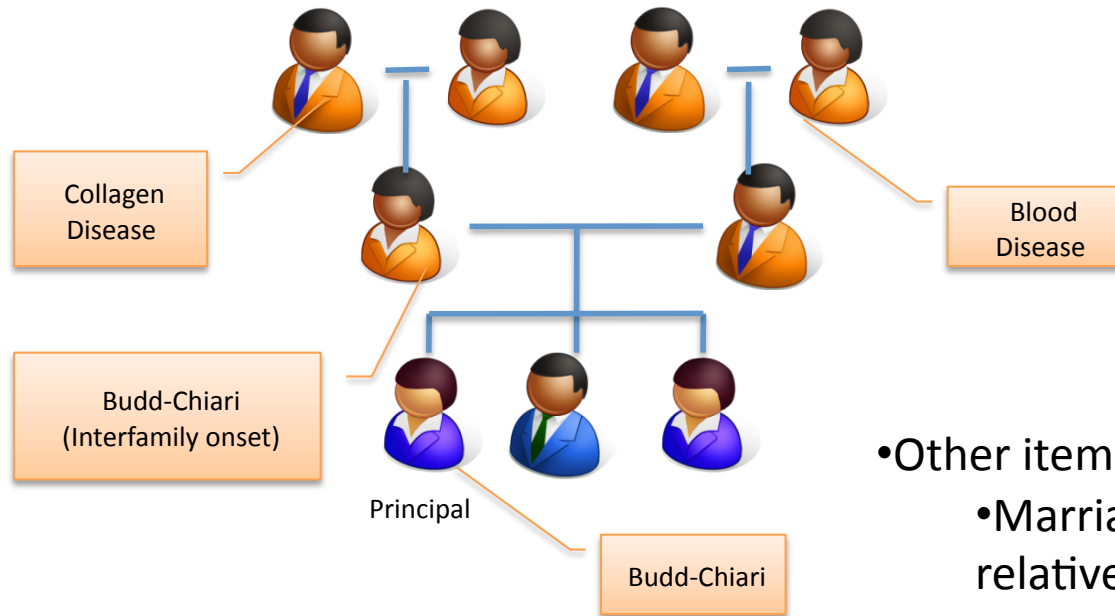
Insurance

- Universality?
 - Public, private, conditions, country
 - Domestic matter?
- Admin entry?
 - Needs for billing procedure

Family History (cont.)

Family History for Budd-Chiari syndrome

- (1) Interfamily onset 1.Yes (Relation:)
2.No
- (2) Collagen Disease 1.Yes (Disease Name: /Relation:) 2.No
- (3) Blood disease 1.Yes (Disease Name: /Relation:) 2.No
- (4) Venous thrombosis 1.Yes (Disease Name: /Relation:) 2.No

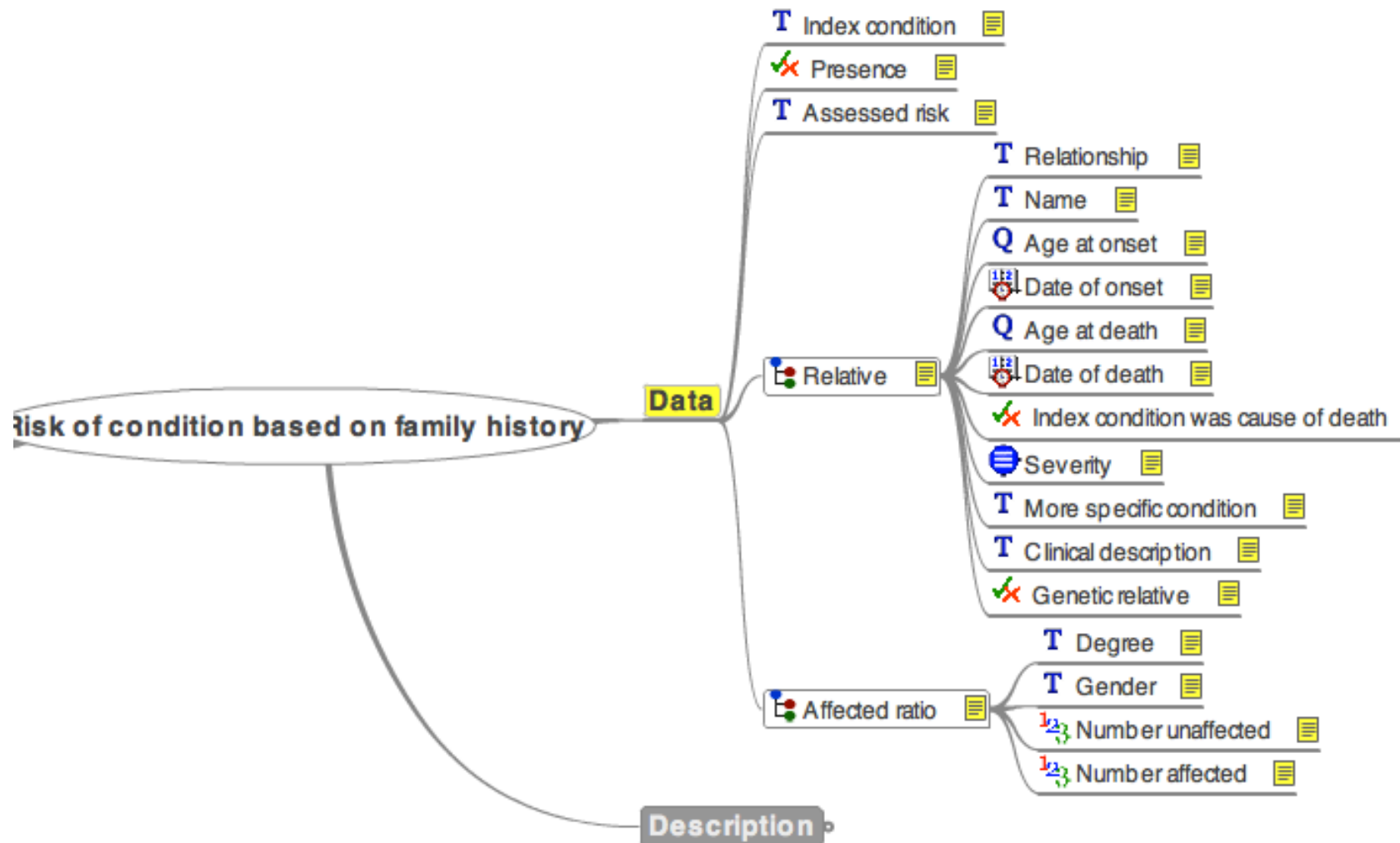


•Other items

- Marriage between relatives
- Born in closed community

- What model will be suitable for describing following family history?

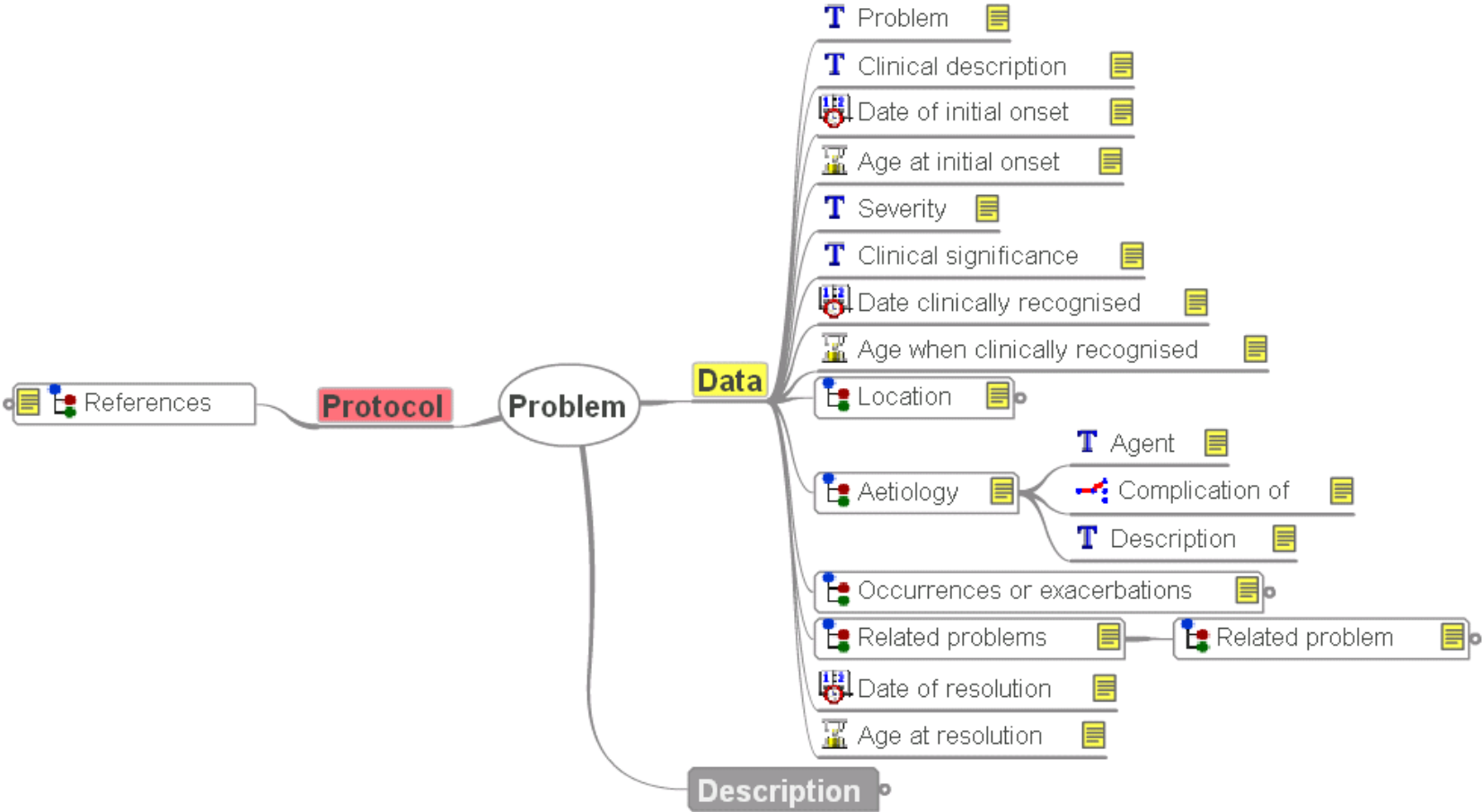
Family History Archetype



Severity

- Many criteria
 - TNM, CTCAE, MGFA, Yahr, UC, Chron
 - Grading?, Staging?
- Compatibility?
 - MG severity(MGFA) vs Parkinson severity(Yahr)
 - Non sense
 - CTCAE/NIH-CTC
 - Partially meaningful

Problem and diagnostic archetype



Modality

- Diagnostic imaging archetype
 - X-Ray, CT, MRI..
 - Detailed condition?
 - Vendor name, Serial No., kVp....
 - Procedure
 - Operation
 - Endoscopy

Terminology

- SNOMED-CT
 - For western countries?
 - Applicable in Asia, Africa?
- Free/Open/Libre terminology?
 - Wikipedia/SNS type terminology
 - Responsibility?

Summary

- Localisation
 - Demographics, Insurance
- For intractable disease
 - General practice <-> Specialised medical service
- Terminology
 - Assurance for Non SNOMED countries