The ROYAL COLLEGE of OPHTHALMOLOGISTS

Clinical Dataset

Uveitis Dataset

The Royal College of Ophthalmologists

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1 Uveitis and the Principles of a Common Dataset

1.1 Introduction

Uveitis describes a complex collection of conditions characterized by intraocular inflammation.[1] As a group, uveitis is a significant cause of blindness worldwide. In the industrialised world it is thought to account for about 10–15% of the cases of total blindness (World Health Organization definition) and up to 20% of legal blindness.[2-3]

In addition to the direct visual impairment that may result from intraocular inflammation, the impact of uveitis is compounded by two important factors: first, many forms of uveitis are associated with significant systemic disease;[4] and second, many of the sight-threatening forms of uveitis require local and/or systemic therapies that are accompanied by significant drug-related morbidity.[5]

The study of the clinical practice of uveitis – both for research and auditing purposes – is challenged by the individual scarcity of most of the constituent syndromes,[6] and a lack of consensus about what a 'gold standard' of optimal care would look like.[7] Valuable data does exist but is usually retrospective, being based on the post-hoc interpretation of case-notes, as seen in the Systemic Immunosuppressive Therapy for Eye Diseases (SITE) Cohort Study.[8] The relatively few number of studies that are prospective are narrow in scope, usually syndrome-specific prospective studies such as in Birdshot Chorioretinopathy.[9] These syndrome-specific studies are valuable in their own right but do not enable inter-syndrome comparisons.

The Royal College of Ophthalmologists has been supporting the development of Datasets for over a decade, with national Datasets for both cataract and diabetic retinopathy, among others. Such Datasets help provide a standardized language for clinical care, and a tool for outcome analysis, clinical audit, revalidation, and research. Although the formation of a Dataset for uveitis is particularly challenging, the future benefits are apparent, with the potential to mark major shifts both in clinical care (including auditing and bench-marking) and the research environment.

1.2 Aims

The overall purpose of this proposal is to provide a Dataset that enables standardization of clinical data collection reflective of routine clinical care and of value for audit and research purposes. Specific aims within this are to provide:

1) a minimum Dataset which is mandatory
2) an extended Dataset which is desirable but not mandatory

It is not within the scope of this work to define a Dataset by which quality of care can be audited for revalidation purposes but it would be anticipated that such a Dataset would be a subset of the minimum dataset, and would take into account the distribution of cases (type

and severity), their therapeutic interventions and their visual outcome over time. Patient reported outcomes would also be desirable but are not currently collected in routine practice for most ophthalmic conditions.

1.3 Principles

1.3.1 General

This dataset ascribes to the core principles elucidated in other RCOphth Datasets[10] namely that:

- The minimum Dataset should be a subset of information routinely collected so as not to add to the demands on busy clinicians.
- Data Elements should only be included if they are identified as being subjects of interest that will be analysed.
- Elements in common with other College Datasets should be congruent; for the purposes of this report we do not describe these in detail but defer to the relevant Dataset.
- The Dataset should be capable of implementation in an electronic medical record.

1.3.2 The Uveitis Minimum Dataset

The minimum Dataset comprises those elements that should be recorded in every case of uveitis, and would be regarded as standard of care for those conditions. The elements included here are those that were considered essential to defining the *type of uveitis*, the *severity of disease*, the *major therapeutic interventions* and the *current status*.

- The *type of uveitis* is defined according to anatomical classification and course (both according to the Standardization of Uveitis Nomenclature recommendations[11]), and aetiological classification (major headings according to the International Uveitis Study Group[1]).
- The *current status* comprises *current visual function* as measured by visual acuity and the *current measures of active inflammation* comprising key measures of active inflammation that have been described according to standardized grades (AC cells, AC flare and vitreous haze).
- The *severity and time-course of uveitis* is assessed by the collection of key measures of active inflammation over time and the presence of potentially sight-threatening complications; the date of the patient's first presentation and the date of first onset of any sight-threatening complications are also recorded.
- The *major therapeutic interventions* comprise significant medical and surgical interventions and their approximate dates.

The Uveitis Minimum Dataset is discussed further below and is provided as Appendix A.

1.3.3 The Uveitis Extended Dataset

The extended Dataset comprises the above data elements plus many additional elements that are relevant to the clinical assessment or treatment pathway in some or all types of uveitis. The extended Dataset is included to provide guidance to electronic medical records developers regarding the inclusion and format of those elements of the clinical record that

are particularly relevant to the care of patients with uveitis. Although it would be ideal to have the extended Dataset recorded in all patients, it is recognized that in routine clinical practice the capture of different elements will be prioritised over others according to their perceived relevance to the uveitis syndrome in question, and thus completion of these elements will be variable. The provision of the extended Dataset does however provide the opportunity for electronic medical record developers to ensure standardization of fields so that in those cases where clinicians *do* perceive their assessment and recording to be important, these data can be aggregated and analysed. It is recognized that even the Extended Dataset is not exhaustive, as the nature of uveitis means that a truly comprehensive Dataset would include an unmanageable number of elements of both ophthalmic and systemic disease, their complications and treatments

The Uveitis Extended Dataset is discussed further below and is provided in Appendix B.

1.4 Specific Challenges

It is acknowledged that the development of a core dataset for uveitis is made more challenging due its complexity, heterogeneity, and limitations in the current system of taxonomy and classification which serve to highlight our imperfect understanding of the aetiology of much uveitis. It is however partly because of these challenges – and the variable ways in which much uveitis is recorded and reported – that a standardized Dataset for uveitis would be of immense value, enabling prospective data gathering in a coherent large-scale manner. There will inevitably be debate as to what characteristics are essential vs desirable, but the priority for the mandatory elements that comprise the core dataset has been to collect those variables that are broadly applicable to all forms of uveitis and reflect the overall care of those patients.

2 The Uveitis Dataset

The minimum Dataset comprises those elements that should be recorded in every case of uveitis, and would be regarded as standard of care for those conditions.

2.1 Type of Uveitis

2.1.1 Anatomical classification of uveitis

Mandatory Data element = Anatomical classification of uveitis according to the Standardization of Uveitis Nomenclature[11] down to ‘subvalue’ level.

Table 1 Anatomical classification of uveitis as per SUN[11]

Value	Definition: *Uveitis in which the primary site of inflammation is*	Subvalue
Anterior	Anterior chamber	
Intermediate	Vitreous	**Pars planitis**
		Non-pars planitis
Posterior	Choroid or retina	**Choroiditis**
		Chorioretinitis
		Retinochoroiditis
		Retinitis
		Neuroretinitis
Panuveitis	Anterior chamber, vitreous and retina or choroid	

Choroiditis, chorioretinitis, retinochoroiditis and retinitis may be subclassified as unifocal, paucifocal, multifocal or diffuse; this is desirable but not mandatory.

2.1.2 Course of uveitis

Mandatory Data element = Course of uveitis according to SUN Descriptors [11].

Table 2 Classification of course of uveitis

Value	Definition
Acute	Episode characterized by sudden onset and limited (ie less than equal to 3 months) duration
Recurrent	Repeated episodes separated by periods of inactivity without treatment of at least 3 months in duration
Chronic	Persistent uveitis (ie more than 3 months duration) with relapse in less than 3 months after discontinuing treatment

These terms are used as per their SUN definitions i.e. duration of attack is defined as limited if it is less than or equal to 3 months, and persistent if it lasts longer than 3 months. Course of uveitis is defined as acute, recurrent or chronic as per their SUN definitions (Table 2). In the extended dataset the onset of uveitis (i.e. sudden vs insidious) is also recorded.

2.1.3 Clinical and Taxonomic classification

Table 3 Clinical classification reflecting taxonomy

Element	Value	Subvalue	Sub-subvalue	Sub-sub-subvalue
Classification by taxonomy	**Infectious**	Bacterial	Mycobacterium tuberculosis (TB)	Ocular only With extraocular involvement
			Treponema pallidum (Syphilis)	Ocular only With extraocular involvement
			Bartonella henselae (Cat-scratch disease)	Ocular only With extraocular involvement
			Borrelia burgdorferi (Lyme disease)	Ocular only With extraocular involvement
			Other bacterial	Specify
		Viral	HSV1	Anterior uveitis Keratouveitis Acute Retinal Necrosis Progressive Outer Retinal Necrosis
			HSV2	Anterior uveitis Keratouveitis Acute Retinal Necrosis Progressive Outer Retinal Necrosis
			VZV	Anterior uveitis Keratouveitis Acute Retinal Necrosis Progressive Outer Retinal Necrosis
			CMV	Anterior uveitis Keratouveitis Posterior uveitis

Element	Value	Subvalue	Sub-subvalue	Sub-sub-subvalue
			Viral syndrome (undifferentiated)	Anterior uveitis Keratouveitis Acute Retinal Necrosis Progressive Outer Retinal Necrosis
			Other viral	Specify
		Fungal	Candida sp.	
			Aspergillus sp.	
			Other fungal	Specify
		Parasitic	Toxoplasma gondii	
			Toxocara canis	
			Onchocerca volvulus	
			Diffuse unilateral subacute necrosis	
			Other parasitic	Specify
		Other		
	Non-Infectious	No systemic disease	Acute/Recurrent Anterior Uveitis	HLA-B27-positive HLA-B27-negative HLA-B27 unknown
			Chronic Anterior Uveitis	
			Fuchs Uveitis Syndrome	
			Intermediate Uveitis	Pars planitis Non-pars planitis
			Acute Posterior Multifocal Placoid Pigment Epitheliopathy	
			Multiple Evanescent White Dot Syndrome	
			Multifocal Choroiditis with Panuveitis	
			Punctate Inner Choroidopathy	
			Ampiginous Choroiditis	
			Birdshot Chorioretinopathy	
			Serpiginous Choroiditis	

Element	Value	Subvalue	Sub-subvalue	Sub-sub-subvalue
			Other non-infectious (no systemic disease)	Specify
		Systemic disease	Ankylosing spondylitis	
			Sarcoidosis	
			Multiple sclerosis	
			Behcet Disease	
			Sympathetic Ophthalmia	
			Vogt-Koyanagi-Harada Disease	
			Tubulointerstitial Nephritis and Uveitis	
			Juvenile idiopathic arthritis	
			Other non-infectious (systemic disease)	Specify
	Masquerade	Neoplastic		Specify
		Non-neoplastic		
	Undifferentiated			

The Clinical Classification of the International Uveitis Study Group[1] provide major clinical headings which to some extent reflects aetiology, at least where this is known. It is recognized that, for many of the 'Non-infectious' group, aetiology is poorly defined particularly for those syndromes not associated with a systemic disease. For this reason we refer to this as 'classification by taxonomy'. The formation of a hierarchical sub-classification beyond that provided by the IUSG is difficult, but is clearly desirable in order to support consistency in EMR systems. The system proposed remains consistent with the IUSG outline and the ongoing work of SUN. A couple of points are worth noting in terms of our proposed hierarchical system for EMR use:

- '*Undifferentiated*' categories: At the point of presentation it may not be possible to accurately classify the type of uveitis using the IUSG system, and therefore '*Undifferentiated*' (rather than idiopathic) may be the most appropriate category. Additionally there are a number of clinically defined infectious syndromes where an infectious aetiology is recognized but the aetiological agent is not fully established at the point of diagnosis, for example Acute Retinal Necrosis. To enable appropriate capture of this type of data we have provided the sub-category of 'Viral Syndrome (Undifferentiated)' with further classification according to the clinical syndromes of Anterior Uveitis, Keratouveitis, Acute Retinal Necrosis and Progressive Outer Retinal Necrosis.

- *Subvalue lists:* 'drop-down' lists are one of the areas where there needs to be a balance between comprehensiveness and usability. Rather than providing exhaustive lists of uveitis entities for each sub-category, we have restricted this to the more common or distinct uveitic diagnoses in each category, with an option of 'Other: please specify' for all those not specifically listed.

It is recognized that most non-infectious syndromes are currently classified by pattern rather than by a detailed understanding of their aetiology, and it is therefore likely that their categorisation may need to be reassessed as knowledge of their pathogenesis improves.

2.2 Current Status

2.2.1 Visual acuity and other measures of visual function

Mandatory Data Element = Visual Acuity

Visual acuity is common to all Datasets and should be standardized. We recommend that Best Corrected LogMAR Visual Acuity for each eye (as measured by ETDRS charts) is used in all cases where possible.

In the extended dataset, if patients have had automated perimetry performed, MD and PSD should also be recorded under investigations.

2.2.2 Current inflammatory activity

2.2.2.1 AC Cells, AC Flare and Vitreous Haze

Mandatory Data Element = AC Cells (0, 0.5, 1, 2, 3 or + according to SUN [11])

Mandatory Data Element = AC Flare (0, 1, 2, 3 or 4 according to SUN [11])

Mandatory Data Element = Vitreous Haze (0, 0.5, 1, 2, 3, 4+ according to SUN/Nussenblatt [11,12])

Severity of uveitis comprises key measures of active inflammation that have been described according to standardized grades by SUN notably AC cells, AC flare and vitreous haze. [11,12] Although it is recognized that AC flare and vitreous are not always recorded by non-experts these are important parameters which should be recorded in all cases of patients with uveitis and thus form part of the minimum Dataset.

2.2.2.2 Summary of Disease Activity

Mandatory Data Element = Summary of Disease Activity

Summary of disease activity provides an overall estimate of inflammatory status. It is described in Table 4. Although the SUN working group suggested definitions of 'worsening' and 'improving' that required a two-step change of AC cells and vitreous haze, this was primarily for use in clinical trials. In everyday clinical practice, the experienced specialist will make an assessment of disease status using all available signs of clinical inflammation, and may regard a lower threshold of change in one or more parameters as significant. This summary of disease activity is therefore meant to be a *subjective overall assessment of active inflammation*, and is not defined in terms of individual components of inflammation. The category of 'In Remission off treatment' equates to the 'In remission' category recommended by SUN.

Table 4 Summary of disease activity

Value	Subvalue	Definition
Active	Worsening	Evidence of increased ocular inflammation
	No change	No apparent change in level of ocular inflammation
	Improving	Evidence of reduced ocular inflammation but still active
Quiescent	Inactivity of recent onset	Disease inactive < 3 months
	In Remission on Treatment	Disease inactive ≥ 3 months but on treatment
	In Remission off Treatment	Disease inactive ≥ 3 months and off treatment

2.2.3 Other quantitative measures

2.2.3.1 Intraocular pressure (IOP)

For each eye:

Mandatory Data Element = IOP (mmHg)

IOP may be impacted directly and indirectly by uveitis (activity, damage or therapy) and may lead to sight loss if abnormally high (glaucoma) or low (hypotony). It is therefore an essential part of the uveitis dataset.

2.3 Time-course and complications of uveitis

2.3.1 Date of onset

For each eye:

Mandatory Data Element = Date of first onset of uveitis

Date of current visit (recorded for all visits) and date of onset of uveitis in each eye are mandatory. It is accepted that date of onset is likely to be an estimate and should be recorded to the nearest month; occasionally it may not be possible to obtain an estimate of the onset of uveitis and for this reason 'not known' should be provided as an alternative entry. Duration of disease is calculated from these parameters where known.

2.3.2 Complications

For each eye:

Mandatory Data Element = Presence of cataract

Mandatory Data Element = Presence of glaucoma or ocular hypertension

Mandatory Data Element = Presence of macular oedema

Mandatory Data Element = Presence of any other sight-threatening complication

Table 5 Presence of potentially sight-threatening complications of uveitis

Element	Value	Subvalue
Cataract	Present	Visually significant Visually insignificant
	Previous	Pseudophakic *(autofill)*
		Aphakic *(autofill)*
	Absent	
Glaucoma/OHT	Present	Glaucoma OHT without glaucomatous optic neuropathy
	Previous	Previous OHT with no ongoing treatment requirement
	Absent	
Macular oedema	Present	Visually significant Visually insignificant
	Previous	
	Absent	

Element	Value	Subvalue
Epiretinal membrane	Present	Visually significant Visually insignificant
	Previous	
	Absent	
Any other visually significant complication	Present	*Please specify*
	Previous	*Please specify*
	Absent	

Key events in the time-course of uveitis are also recorded, notably the date of first presentation of uveitis and the presence of any sight-threatening complications; the presence or absence of cataract, glaucoma, macular oedema and epiretinal membrane and whether they are visually significant (Table 5) must be recorded in all cases. Any other potentially sight-threatening complications should be listed under 'Any other visually significant complication'.

In the **extended Dataset**, the dates when these complications were first noted are recorded to the nearest month and may be estimated where the precise date is not known.

For each eye (where presence of sight-threatening complication has been noted):

Extended Data Element = Date cataract first noted

Extended Data Element = Date glaucoma first noted

Extended Data Element = Date macular oedema first noted

Extended Data Element = Date epiretinal membrane first noted

Extended Data Element = Date other specified sight-threatening complication first noted

2.4 Major therapeutic interventions

It is essential that both medical and surgical therapies directly related to the uveitis or its complications are recorded. It is recognized that most EMR systems will gather this data routinely (and indeed more comprehensively than outlined below) as part of their standard medical therapy interface pages. *It is not expected that the clinician would be required to re-enter any of this data*. These variables are however highlighted to support what would be expected to be available in terms of data extraction as they highlight key therapeutic interventions that are of ongoing relevance to the patient.

2.4.1 Relevant medical therapy

For each eye:

Mandatory Data Element = Current topical corticosteroid therapy

Mandatory Data Element = Local therapy within the last two years

Not specific to eye:

Mandatory Data Element = Current oral corticosteroid

Mandatory Data Element = Intravenous corticosteroid therapy within the last three months

Mandatory Data Element = Intramuscular corticosteroid therapy within the last three months

Mandatory Data Element = Current other immunosuppressants

Table 6 Current medical therapy for uveitis

Element	Value	Subvalue
For each eye		
Topical corticosteroid therapy (current)	Present	*Specify drug, dose, frequency*
	Absent	
Local therapy within the last two years	Present	*Specify drug, route and date*
Per patient		
Oral corticosteroid (current)	Present	Reducing course *(specify current dose)*
		Maintenance dose *(specify dose)*
	Absent	
Intravenous corticosteroid (≤ 3 mths)	Present	*Specify drug, dose, date*
	Absent	
Intramuscular corticosteroid (≤ 3 mths)	Present	*Specify drug, dose, date*
	Absent	
Other immunosuppressant (current)	Present	*Specify drug, dose, route, frequency*
	Absent	

In the **extended Dataset**, any additional *active* or *significant previous* ocular treatments should be recorded. In the *active* group, common additional medications of particular interest are the topical therapies for glaucoma. In the *significant previous* treatments group, medications of particular interest are previous systemic immunosuppressant drugs, local therapies prior to two years earlier, and intravenous or intramuscular corticosteroid prior to three months earlier. Adverse drug reactions which may influence future treatment decisions in relation to uveitis, such as corticosteroid-induced ocular hypertension or hepatotoxicity due to a specific immunosuppressant, are also recorded in the extended dataset.

2.4.2 Relevant surgical therapy

For each eye:

Mandatory Data Element = Cataract surgery or related procedures

Mandatory Data Element = Glaucoma surgery or related procedures

Mandatory Data Element = Vitreo-retinal surgery or related procedures

Mandatory Data Element = Any other ocular surgery

Table 7 Relevant surgical therapy

Element	Value	Subvalue
Cataract surgery	Present	*Please specify operation(s) and date(s)*
	Absent	
Glaucoma surgery	Present	*Please specify operation(s) and date(s)*
	Absent	
Vitreo-retinal surgery	Present	*Please specify operation(s) and date(s)*
	Absent	
Any other ocular surgery	Present	*Please specify operation(s) and date(s)*

The dates when these interventions were performed are recorded to the nearest month and may be estimated where the precise date is not known. Related procedures include interventions such as YAG posterior capsulotomy following cataract surgery.

2.5 Investigations

Although not included in the minimum dataset, the extended dataset includes selected investigations that are important either for diagnosis or monitoring of disease. The gathering of this data is encouraged both for assessing the current usage of these tests for particular uveitic syndromes and may provide some estimate of the diagnostic utility of these investigations in this context. Quantitative data from common serial assessments that are used for disease monitoring, such as central macular thickness as measured by optical coherence tomography, is also recorded.

3 References

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A1. Classification of uveitis

A1.1 Classification by anatomical category

Element	Value	Definition: Uveitis in which the primary site of inflammation is	Subvalue
Classification by anatomical category	**Anterior**	Anterior chamber	
	Intermediate	Vitreous	**Pars planitis**
			Non-pars planitis
	Posterior	Choroid or retina	**Choroiditis**
			Chorioretinitis
			Retinochoroiditis
			Retinitis
			Neuroretinitis
	Panuveitis	Anterior chamber, vitreous and retina or choroid	

A1.2 Classification by course

Element	Value	Definition
Classification by course	**Acute**	Episode characterized by sudden onset and limited (ie less than 3 months) duration
	Recurrent	Repeated episodes separated by periods of inactivity without treatment of at least 3 months in duration
	Chronic	Persistent uveitis with relapse in less than 3 months after discontinuing treatment

A1.3 Classification by taxonomy

Element	Value	Subvalue	Sub-subvalue	Sub-sub-subvalue
Taxonomy	**Infectious**	Bacterial	Mycobacterium tuberculosis (TB)	Ocular only With extraocular involvement
			Treponema pallidum (Syphilis)	Ocular only With extraocular involvement
			Bartonella henselae (Cat-scratch disease)	Ocular only With extraocular involvement
			Borrelia burgdorferi (Lyme disease)	Ocular only With extraocular involvement
			Other bacterial	Specify
		Viral	HSV1	Anterior uveitis Keratouveitis Acute Retinal Necrosis Progressive Outer Retinal Necrosis
			HSV2	Anterior uveitis Keratouveitis Acute Retinal Necrosis Progressive Outer Retinal Necrosis
			VZV	Anterior uveitis Keratouveitis Acute Retinal Necrosis Progressive Outer Retinal Necrosis
			CMV	Anterior uveitis Keratouveitis Posterior uveitis
			Viral syndrome (undifferentiated)	Anterior uveitis Keratouveitis Acute Retinal Necrosis Progressive Outer Retinal Necrosis

Element	Value	Subvalue	Sub-subvalue	Sub-sub-subvalue
			Other viral	Specify
		Fungal	Candida sp.	
			Aspergillus sp.	
			Other fungal	
		Parasitic	Toxoplasma gondii	
			Toxocara canis	
			Onchocerca volvulus	
			Diffuse unilateral subacute necrosis	
			Other parasitic	Specify
		Other		Specify
	Non-Infectious	No systemic disease	Acute/Recurrent Anterior Uveitis	HLA-B27-positive HLA-B27-negative HLA-B27 unknown
			Chronic Anterior Uveitis	
			Fuchs Uveitis Syndrome	
			Intermediate Uveitis	Pars planitis Non-pars planitis
			Acute Posterior Multifocal Placoid Pigment Epitheliopathy	
			Multiple Evanescent White Dot Syndrome	
			Multifocal Choroiditis with Panuveitis	
			Punctate Inner Choroidopathy	
			Ampiginous Choroiditis	
			Birdshot Chorioretinopathy	

Element	Value	Subvalue	Sub-subvalue	Sub-sub-subvalue
			Serpiginous Choroiditis	
			Other non-infectious (no systemic disease)	Specify
		Systemic disease	Ankylosing spondylitis	
			Sarcoidosis	
			Multiple sclerosis	
			Behcet Disease	
			Sympathetic Ophthalmia	
			Vogt-Koyanagi-Harada Disease	
			Tubulointerstitial Nephritis and Uveitis	
			Juvenile idiopathic arthritis	
			Other non-infectious (systemic disease)	Specify
	Masquerade	Neoplastic		Specify
		Non-neoplastic		Specify
	Undifferentiated			

A2. Current status

A2.1 Visual acuity for each eye

Element	Value
Visual acuity	
Best corrected	LogMAR value

A2.2 Current inflammatory activity for each eye

Element	Value
AC cells	0-4 as per SUN
AC flare	0-4 as per SUN
Vitreous haze	0-4 as per SUN/Nussenblatt

A2.3 Intraocular pressure

Element	Value
Intraocular pressure	Numerical (mmHg)

A2.4 Summary of disease activity for each eye

Element	Value	Subvalue	Definition
Summary of disease activity	**Active**	Worsening	Evidence of increased ocular inflammation
		No change	No apparent change in level of ocular inflammation
		Improving	Evidence of reduced ocular inflammation but still active
	Quiescent	Inactivity of recent onset	Disease inactive < 3 months
		In Remission on Treatment	Disease inactive $\geq$ 3 months but on treatment
		In Remission off Treatment	Disease inactive $\geq$ 3 months and off treatment

A3. Time-course and complications of uveitis

A3.1 Time-course

Element	Value	Subvalue
Date of first onset of uveitis (per eye)	*Specify*	
Date of current visit	*Specify*	
Duration of disease since first onset	*Calculated*	

A3.2 Complications for each eye

Element	Value	Subvalue
Cataract	Present	Visually significant Visually insignificant
	Previous	Pseudophakic *(autofill)*
		Aphakic *(autofill)*
	Absent	
Glaucoma/OHT	Present	Glaucoma OHT without glaucomatous optic neuropathy
	Previous	Previous OHT with no ongoing treatment requirement
	Absent	
Macular oedema	Present	Visually significant Visually insignificant
	Previous	
	Absent	
Epiretinal membrane	Present	Visually significant Visually insignificant
	Previous	
	Absent	

Element	Value	Subvalue
Any other visually significant complication	Present	*Please specify*
	Previous	*Please specify*
	Absent	

A4. Major therapeutic interventions

A4.1 Relevant Medical Therapies

Element	Value	Subvalue
For each eye		
Topical corticosteroid therapy (current)	Present	*Specify drug, dose, frequency*
	Absent	
Local therapy within the last two years	Present	*Specify drug, route and date*
Per patient		
Oral corticosteroid (current)	Present	Reducing course *(specify current dose)*
		Maintenance dose *(specify dose)*
	Absent	
Intravenous corticosteroid (≤ 3 mths)	Present	*Specify drug, dose, date*
	Absent	
Intramuscular corticosteroid (≤ 3 mths)	Present	*Specify drug, dose, date*
	Absent	
Other immunosuppressant (current)	Present	*Specify drug, dose, route, frequency*
	Absent	

A4.2 Relevant surgical therapies for each eye

Element	Value	Subvalue
Cataract surgery	Present	*Please specify operation(s) and date(s)*
	Absent	
Glaucoma surgery	Present	*Please specify operation(s) and date(s)*
	Absent	
Epiretinal membrane surgery	Present	*Please specify operation(s) and date(s)*
	Absent	
Any other ocular surgery	Present	*Please specify operation(s) and date(s)*

Appendix B. Extended dataset for clinical features:

The extended dataset contains all features of the minimum dataset but greater detail in terms of clinical features, course of disease and interventions. For clarity those **elements** that are contained within both sets (i.e. mandatory data elements) are marked in **bold**.

B1. Classification of uveitis

B1.1 Classification by anatomical category

Element	Value	Definition: Uveitis in which the primary site of inflammation is	Subvalue	Sub-subvalue
Classification by anatomical category	Anterior	Anterior chamber		
	Intermediate	Vitreous	Pars planitis	
			Non-pars planitis	
	Posterior	Choroid or retina	Choroiditis	Unifocal Paucifocal Multifocal Diffuse
			Chorioretinitis	Unifocal Paucifocal Multifocal Diffuse
			Retinochoroiditis	Unifocal Paucifocal Multifocal Diffuse
			Retinitis	Unifocal Paucifocal Multifocal Diffuse
			Neuroretinitis	
	Panuveitis	Anterior chamber, vitreous and retina or choroid		

B1.2 Classification by course

Element	Value	Definition
Classification by course	Acute	Episode characterized by sudden onset and limited (ie less than 3 months) duration
	Recurrent	Repeated episodes separated by periods of inactivity without treatment of at least 3 months in duration
	Chronic	Persistent uveitis with relapse in less than 3 months after discontinuing treatment

Element	Value
Descriptor of uveitis onset	Sudden
	Insidious

B1.3 Classification by aetiology/clinical syndrome

Element	Value	Subvalue	Sub-subvalue	Sub-sub-subvalue
Classification by Taxonomy	**Infectious**	Bacterial	Mycobacterium tuberculosis (TB)	Ocular only With extraocular involvement
			Treponema pallidum (Syphilis)	Ocular only With extraocular involvement
			Bartonella henselae (Cat-scratch disease)	Ocular only With extraocular involvement
			Borrelia burgdorferi (Lyme disease)	Ocular only With extraocular involvement
			Other bacterial	Specify
		Viral	HSV1	Anterior uveitis Keratouveitis Acute Retinal Necrosis Progressive Outer Retinal Necrosis
			HSV2	Anterior uveitis Keratouveitis Acute Retinal Necrosis Progressive Outer Retinal Necrosis
			VZV	Anterior uveitis Keratouveitis Acute Retinal Necrosis Progressive Outer Retinal Necrosis
			CMV	Anterior uveitis Keratouveitis Posterior uveitis
			Viral syndrome (undifferentiated)	Anterior uveitis Keratouveitis Acute Retinal Necrosis Progressive Outer Retinal Necrosis

Element	Value	Subvalue	Sub-subvalue	Sub-sub-subvalue
			Other viral	Specify
		Fungal	Candida sp.	
			Aspergillus sp.	
			Other fungal	Specify
		Parasitic	Toxoplasma gondii	
			Toxocara canis	
			Onchocerca volvulus	
			Diffuse unilateral subacute necrosis	
			Other parasitic	Specify
		Other		Specify
	Non-Infectious	No systemic disease	Acute/Recurrent Anterior Uveitis	HLA-B27-positive HLA-B27-negative HLA-B27 unknown
			Chronic Anterior Uveitis	
			Fuchs Uveitis Syndrome	
			Intermediate Uveitis	Pars planitis Non-pars planitis
			Acute Posterior Multifocal Placoid Pigment Epitheliopathy	
			Multiple Evanescent White Dot Syndrome	
			Multifocal Choroiditis with Panuveitis	
			Punctate Inner Choroidopathy	
			Ampiginous Choroiditis	
			Birdshot Chorioretinopathy	

Element	Value	Subvalue	Sub-subvalue	Sub-sub-subvalue
			Serpiginous Choroiditis	
			Other non-infectious (no systemic disease)	Specify
		Systemic disease	Ankylosing spondylitis	
			Sarcoidosis	
			Multiple sclerosis	
			Behcet Disease	
			Sympathetic Ophthalmia	
			Vogt-Koyanagi-Harada Disease	
			Tubulointerstitial Nephritis and Uveitis	
			Juvenile idiopathic arthritis	
			Other non-infectious (systemic disease)	Specify
	Masquerade	Neoplastic		Specify
		Non-neoplastic		Specify
	Undifferentiated			

B2. Current status

B2.1 Visual function

Element	Value	Subvalue
Visual acuity		
VA - Unaided	LogMAR value	
VA - Best corrected	LogMAR value	
VA - Pin-hole	LogMAR value	
Colour vision	Number of Ishihara plates (x/y)	
RAPD	Present	
	Absent	

B2.2 Current inflammatory activity and other clinical features for each eye

Symptoms

Element	Value	Subvalue
Photophobia	Present	
	Absent	
Other eye pain	Present	
	Absent	
Blurred vision	Present	
	Absent	
Floaters	Present	
	Absent	
Photopsia	Present	
	Absent	
Blind-spots	Present	
	Absent	
Distortion	Present	
	Absent	
Other visual symptoms	Present	*Please specify*
	Absent	
Other non-visual ocular symptoms	Present	*Please specify*
	Absent	

Conjunctiva and sclera

Element	Value	Subvalue	Sub-subvalue
Conjunctival injection	Present	Diffuse	Severity (0-4)
		Sectoral	Severity (0-4)
		Circumlimbal	Severity (0-4)
	Absent		
Episcleritis	Present	Nodular	
		Non-nodular	Diffuse Sectoral
	Absent		
Anterior Scleritis	Present	Non-necrotising	Diffuse Nodular
		Necrotising	With evident inflammation Without evident inflammation (scleromacia perforans-type)
	Absent		

Cornea

Element	Value	Subvalue	Sub-subvalue
Keratic precipitates	Present	Regular	Diffuse Inferior Focal
		Granulomatous	Diffuse Inferior Focal
		Stellate	Diffuse Inferior Focal
Band keratopathy	Present	Involves visual axis	
		Not involving visual axis	
	Absent		
	Not done		

Element	Value	Subvalue	Sub-subvalue
Corneal oedema	Present	*Specify level*	
	Absent		
Pachymetry	Done	*Specify CCT in microns*	

Anterior Chamber

Element	Value	Subvalue	Sub-subvalue
AC cells	0-4 as per SUN		
AC flare	0-4 as per SUN		
Hypopyon	Present	*Specify height in mm*	
	Absent		
Fibrin	Present		
	Absent		
Hyphaema	Present	*Specify height in mm*	
	Absent		

Intraocular pressure

Element	Value	Subvalue	Sub-subvalue
Intraocular pressure	0-100 (mmHg)		

Iris

Element	Value	Subvalue	Sub-subvalue
Nodules	Present	Koeppe nodules	
		Busacca nodules	
		Angle nodules	
		Mixed nodules	
	Absent		
Posterior synechiae	Present	*Specify degrees of PS (0-360')*	
	Absent		
Peripheral anterior synechiae	Present	*Specify degrees of PS (0-360')*	

Element	Value	Subvalue	Sub-subvalue
	Absent		
Transillumination of iris (disease-related)	Present	Diffuse	
		Sectoral	
		Radial	
	Absent		
Surgical/traumatic iris defect	Present	Surgical iridectomy	
		Laser iridotomy	
		Traumatic	
	Absent		
Neovascularistaion	Present		
	Absent		
Gonioscopy	*Specify angle conformation according to a recognized grading system*		

Lens

Element	Value	Subvalue	Sub-subvalue
Phakic status	Clear crystalline lens		
	Cataract	Visually significant	Nuclear Cortical PSCLO Other
		Not visually significant	
	Pseudophakic	PCIOL in bag	
		PCIOL in sulcus	
		ACIOL	
		Sutured IOL	
	Aphakic		

Element	Value	Subvalue	Sub-subvalue
Posterior capsule status (where relevant)	Clear		
	PCO	Visually significant	
		Not visually significant	
Anterior capsule status (where relevant)	Normal		
	Anterior capsular phimosis		
Lens stability	Central		
	Subluxed		

Vitreous

Element	Value	Subvalue	Sub-subvalue
Vitreous haze	0-4 as per SUN		
Vitreous cells	Present		
	Absent		
Vitreous snowballs	Present		
	Absent		
Pars plana exudates	Present		
	Absent		
Vitreous haemorrhage	Present		
	Absent		
Posterior vitreous detachment	Present		
	Absent		

Fundus - Macula

Element	Value	Subvalue	Sub-subvalue
Macular oedema	Present		
	Absent		

Element	Value	Subvalue	Sub-subvalue
Other macular pathology	Present	Epiretinal membrane	
		CNV membrane	
		Scar	
		Atrophy	
		Other	*Specify*
	Absent		

Fundus – Choroidal pathology

Element	Value	Subvalue	Sub-subvalue
Choroiditis	Present	Unifocal	
		Paucifocal	
		Multifocal	
	Absent		
Other choroidal lesions	Present	*Specify*	
	Absent		

Fundus – Retinal pathology

Element	Value	Subvalue	Sub-subvalue	Sub-sub-subvalue
Retinitis	Present	Unifocal	*Specify location*	
		Paucifocal		
		Multifocal		
		Diffuse		
	Absent			
Retinal detachment	Present	Serous	Macula-involving	Unifocal Multifocal
			Not macula-involving	Unifocal Multifocal
		Rhegmatogenous	Macula-involving	
			Not macula-involving	
		Tractional	Macula-involving	

Element	Value	Subvalue	Sub-subvalue	Sub-sub-subvalue
			Not macula-involving	
	Absent			
Other retinal pathology	Present	*Specify nature and location*		
	Absent			

Fundus – Retinovascular pathology

Element	Value	Subvalue	Sub-subvalue
Retinal haemorrhages	Present	*Specify location*	
	Absent		
Cotton wool spots	Present	*Specify location*	
	Absent		
Roth spots	Present	*Specify location*	
	Absent		
Vasculitis	Present	Arterial	Active - Focal Active – Diffuse Inactive eg sheathing
		Venous	Active - Focal Active – Diffuse Inactive eg sheathing
		Mixed	Active - Focal Active – Diffuse Inactive eg sheathing
	Absent		
Retinal vein occlusion	Present	BRVO	*Specify location(s)*
		HRVO	Superior Inferior
		CRVO	
		Papillophlebitis	
	Absent		
Retinal arteriole occlusion	Present	BRAO	*Specify location(s)*

Element	Value	Subvalue	Sub-subvalue
		CRAO	
	Absent		
Neovascularisation	Present	NVD	*Specify location(s)*
		NVE	*Specify location(s)*
	Absent		

Optic disc

Element	Value	Subvalue	Sub-subvalue
Cup-Disc Ratio	0-1.0		
Features suspicious of glaucoma	Present	*Specify features of concern*	
	Absent		
Disc colour	Normal		
	Hyperaemic		
	Pallor		
Disc margin	Normal		
	Disc oedema		
	Other	*Specify*	
Disc haemorrhages	Present		
	Absent		
Disc vessels	Normal		
	Disc collaterals		
	NVD		

B2.3 Summary of disease activity for each eye

Element	Value	Subvalue	Definition
Summary of disease activity	Active	Worsening	Evidence of increased ocular inflammation
		No change	No apparent change in level of ocular inflammation

Element	Value	Subvalue	Definition
		Improving	Evidence of reduced ocular inflammation but still active
	Quiescent	Inactivity of recent onset	Disease inactive < 3 months
		In Remission on Treatment	Disease inactive ≥ 3 months but on treatment
		In Remission off Treatment	Disease inactive ≥ 3 months and off treatment

B3. Time-course and complications of uveitis

B3.1 Time-course

Element	Value	Subvalue
Date of first onset of uveitis (per eye)	*Specify*	
Date of current visit	*Specify*	
Duration of disease since first onset	*Calculated*	

B3.2 Complications for each eye

Element	**Value**	**Subvalue**
Cataract	Present	Visually significant Visually insignificant
	Previous	Pseudophakic *(autofill)*
		Aphakic *(autofill)*
	Absent	
Glaucoma/OHT	Present	Glaucoma OHT without glaucomatous optic neuropathy

Element	Value	Subvalue
	Previous	Previous OHT with no ongoing treatment requirement
	Absent	
Macular oedema	Present	Visually significant Visually insignificant
	Previous	
	Absent	
Epiretinal membrane	Present	Visually significant Visually insignificant
	Previous	
	Absent	
Any other visually significant complication	Present	*Please specify*
	Previous	*Please specify*
	Absent	

B4. Major therapeutic interventions

B4.1.1 Relevant Medical Therapies

Element	Value	Subvalue
For each eye		
Topical corticosteroid therapy (current)	Present	*Specify drug, dose, frequency*
	Absent	
Other topical therapies (current)	Present	*Specify drug, dose, frequency*
	Absent	
Local therapy within the last two years	Present	*Specify drug, route and date*
	Absent	

Element	Value	Subvalue
Local therapy prior to two years previously	Present	*Specify drug, route and date*
Absent		
Per patient		
Oral corticosteroid (current)	Present	Reducing course *(specify current dose)*
		Maintenance dose *(specify dose)*
	Absent	
Oral corticosteroid (previous)	Present	*Specify dose-ranges and dates of episodes of treatment*
	Absent	
Intravenous corticosteroid (within last 3 months)	Present	*Specify drug, dose, date*
	Absent	
Intravenous corticosteroid (prior to 3 months previously)	Present	*Specify drug, dose, date*
	Absent	
Intramuscular Corticosteroid (within last 3 months)	Present	*Specify drug, dose, date*
	Absent	
Intramuscular Corticosteroid (prior to 3 months previously)	Present	*Specify drug, dose, date*
	Absent	
Other immunosuppressant (current)	Present	*Specify drug, dose, route, frequency*
	Absent	
Other immunosuppressant (previous)	Present	*Specify dose-ranges, dates of episodes of treatment, and reason for cessation*

Element	Value	Subvalue
	Absent	
Other relevant medical therapies (current)	Present	*Specify drug, dose, route, frequency*
	Absent	

B4.1.2 Significant drug-related adverse events affecting future treatment

Element	**Value**	**Subvalue**	**Sub-subvalue**
Corticosteroid-induced ocular hypertension	Present	Definite	*Please specify drug, dose, date and complication*
		Probable	*Please specify drug, dose, date and complication*
		Possible	*Please specify drug, dose, date and complication*
	Absent		
Other drug reaction requiring cessation or future avoidance	Present	Definite	*Please specify drug, dose, date and complication*
		Probable	*Please specify drug, dose, date and complication*
		Possible	*Please specify drug, dose, date and complication*
	Absent		

B4.2 Relevant surgical therapies for each eye

Element	**Value**	**Subvalue**
Cataract surgery	Present	*Please specify operation(s) and date(s)*
	Absent	
Glaucoma surgery	Present	*Please specify operation(s) and date(s)*

Element	Value	Subvalue
	Absent	
Vitreo-retinal surgery	Present	*Please specify operation(s) and date(s)*
	Absent	
Any other ocular surgery	Present	*Please specify operation(s) and date(s)*

B5. Investigations

B5.1 Quantitative data from common serial assessments for each eye

Element	Value
Central macular thickness on OCT	*Numerical*
MD on 24-2 automated perimetry	*Numerical*
PSD on 24-2 automated perimetry	*Numerical*

B5.2 Genotypic data

Element	Value
HLA-A29	Positive
	Negative
	Not tested
HLA-B27	Positive
	Negative
	Not tested
HLA-B51	Positive
	Negative
	Not tested
HLA-DR4	Positive
	Negative
	Not tested
Other genotypic data	*Specify*

B5.3 Other investigations

Element	Value	Subvalue	Sub-subvalue
ACE	Positive		
	Negative		
	Not tested		
Syphilis serology	Positive		
	Negative		
	Not tested		
Interferon gamma release assay	Positive		
	Negative		
	Inconclusive		
	Not tested		
Mantoux	Positive		
	Equivocal		
	Negative		
	Not tested		
CXR	Suggestive of pathology related to uveitis	*Specify*	
	Abnormal but unrelated to uveitis		
	Normal		
	Not tested		
CT Thorax	Suggestive of pathology related to uveitis	*Specify*	
	Abnormal but unrelated to uveitis		
	Normal		
	Not tested		
MRI Brain	Suggestive of pathology related to uveitis	*Specify*	

Element	Value	Subvalue	Sub-subvalue
	Abnormal but unrelated to uveitis		
	Normal		
	Not tested		
Aqueous humour	PCR	PCR positive	*Specify organism*
		PCR negative	
		PCR not done	
	Other findings	*Specify*	
Vitreous humour	PCR	PCR positive	*Specify organism*
		PCR negative	
		PCR not done	
	Other findings	*Specify*	
Other relevant investigations	*Specify*		