CF ANNUAL ASSESSMENT

Noah's Ark
Children's Hospital for Wales
Ysbyty Plant Cymru

ADDRESSOGRAPH

Date of assessment:

❑ Full care ❑ Llantrisant ❑ Mountain Ash
❑ Shared care ❑ Abergavenny ❑ Hereford
❑ Bridgend ❑ Newport
❑ Carmarthen ❑ Swansea

❑ Face to face ❑ Virtual

Dictated	__________
Typed	__________
Returned	__________
UK CF Registry	__________
Database	__________
Radiology	__________
E-mailed	__________
Posted	__________

DIETITIAN

CORE INFORMATION

Height __________ cm ________ centile Weight __________ kg ________ centile BMI __________ kg/m² ________centile

Oxygen therapy since last annual review: ❑ Yes ❑ No ❑ Not known
If yes: ❑ Continuously ❑ Nocturnal and/or with exertion ❑ During exacerbation ❑ PRN

Has patient received since last annual review: Influenza vaccination ❑ Yes ❑ No ❑ Not known
Pneumococcal vaccination ❑ Yes ❑ No ❑ Not known

Has patient participated in any clinical drug trial since last annual review: ❑ Yes ❑ No ❑ Not known

If yes please specify ____

Has patient participated in any clinical study other than a drug study since last annual review: ❑ Yes ❑ No ❑Not known

If yes please specify ____

ADMISSIONS & IVS

Courses of oral antibiotics in last year __________

IV hospital admissions since last visit (number) __________

Start date	End date	Total days	Admission reason	
			❑ Pulmonary exacerbation ❑ Planned IVs ❑ Pseudomonas aeruginosa eradication	❑ Induction of NTM ❑ Other If other please specify:
			❑ Pulmonary exacerbation ❑ Planned IVs ❑ Pseudomonas aeruginosa eradication	❑ Induction of NTM ❑ Other If other please specify:
			❑ Pulmonary exacerbation ❑ Planned IVs ❑ Pseudomonas aeruginosa eradication	❑ Induction of NTM ❑ Other If other please specify:
			❑ Pulmonary exacerbation ❑ Planned IVs ❑ Pseudomonas aeruginosa eradication	❑ Induction of NTM ❑ Other If other please specify:

Home IVs since last visit (number) __________

Start date	End date	Total days	Reason	
			❑ Pulmonary exacerbation ❑ Planned IVs ❑ Sinus infection	❑ Not known ❑ Other If other please specify:
			❑ Pulmonary exacerbation ❑ Planned IVs ❑ Sinus infection	❑ Not known ❑ Other If other please specify:

Non-IV hospital admissions since last visit ___________

Start date	End date	Total days	Admission reason	
			❑ Non exacerbation pulmonary complication ❑ GI complications ❑ Transplant related ❑ Non transplant surgery ❑ Liver	❑ Bowel ❑ Diabetes ❑ Not known ❑ Other If other please specify:
			❑ Non exacerbation pulmonary complication ❑ GI complications ❑ Transplant related ❑ Non transplant surgery ❑ Liver	❑ Bowel ❑ Diabetes ❑ Not known ❑ Other If other please specify:

PHYSIOTHERAPIST

INVESTIGATIONS

FEV_1 raw value ___________ (l) ❑ Not measured

FEV_1 % predicted ___________

FEF25-75 raw value ___________ (l/s) ❑ Not measured

FEF25-75 % predicted ___________

FVC raw value ___________ (l) ❑ Not measured

FVC % predicted ___________

Best FEV_1 raw value ___________ (l) ❑ Not measured

Best FEV_1 % predicted ________________________

Height ________________________

Weight ________________________

Date ________________________

Oxygen saturations ___________ (%)

Range ________________________

Faecal elastase ___________ (mg/g) ❑ Not known/not done

NURSE

Patient has been screened for CFRD ❑ Yes ❑ No ❑ Prior CFRD Diagnosis ❑ Not known

HbA1c ___________ (mmol/ml) Random blood glucose ___________ (mmol/l)

Fasting blood glucose ___________ (mmol/l) Oral glucose tolerance 2 hour post ___________ (mmol/l)

Continuous glucose monitoring result ❑ Normal ❑ Abnormal ❑ CFRD ❑ Not done

DEXA scan performed: ❑ Normal ❑ Abnormal ❑ Not done ❑ Not known

If done: Date: ___________ DEXA scan lumbar spine <20 years of age (BMAD z-score) ___________

DEXA scan femoral neck <20 years of age (BMAD z-score) ___________

Chest x-ray result: ❑ No change ❑ New changes ❑ Done but result not known ❑ Not done

Comments:

Liver ultrasound scan performed: ❑ Yes ❑ No ❑ Not known If yes: ❑ Normal ❑ Abnormal

Liver appearance: ❑ Normal ❑ Abnormal Spleen size ___________ cm

Portal vein flow rate: ❑ Normal ❑ Abnormal

Comments:

NURSE

Laboratory liver enzymes done: ❑ Yes ❑ No ❑ Not known Date of blood tests ______________________________

ALT __________ ❑ Normal ❑ >1 to <3 x ULN ❑ >3 to >5 x ULN ❑ >5 to <8 x ULN ❑ >8 x ULN

AST __________ ❑ Normal ❑ >1 to <3 x ULN ❑ >3 to >5 x ULN ❑ >5 to <8 x ULN ❑ >8 x ULN

GGT __________ ❑ Normal ❑ >1 to <3 x ULN ❑ >3 to >5 x ULN ❑ >5 to <8 x ULN ❑ >8 x ULN

ALP __________ ❑ Normal ❑ >1 to <2 x ULN ❑ >2 x ULN

Total Bilirubin __________ ❑ Normal ❑ >1 to <2 x ULN ❑ >2 x ULN

Albumin __________

Haemoglobin __________

White blood cells __________

Eosinophils __________

Neutrophils __________

Platelets __________

Prothrombin time __________

CRP __________

Sodium __________

Potassium __________

Urea __________

Serum creatinine __________ (mmol/dl) ❑ Not done

Vitamin B12 __________

Retinol (Vit A) __________

Tocopherol (Vit E) __________

Vitamin D __________

Adjusted Calcium __________

Phosphate __________

IgG __________

IgM __________

IgA __________

IgE __________

Aspergillus fumigatus IgE __________

Aspergillus fumigatus IgG __________

Aspergillus fumigatus cultured: ❑ Yes ❑ No

CHRONIC MEDICATIONS Has this patient had any chronic medications: ❑ Yes ❑ No

ANTIBIOTICS	DOSAGE	FREQUENCY	START DATE	END DATE (OR N/A)
GASTROINTESTINAL	**DOSAGE**	**FREQUENCY**	**START DATE**	**END DATE (OR N/A)**

ENDOCRINE	DOSAGE	FREQUENCY	START DATE	END DATE (OR N/A)
VITAMINS	**DOSAGE**	**FREQUENCY**	**START DATE**	**END DATE (OR N/A)**
INHALERS	**DOSAGE**	**FREQUENCY**	**START DATE**	**END DATE (OR N/A)**
PHYSIOTHERAPY	**DOSAGE**	**FREQUENCY**	**START DATE**	**END DATE (OR N/A)**
OTHER	**DOSAGE**	**FREQUENCY**	**START DATE**	**END DATE (OR N/A)**

CFTR MODULATORS

KAFTRIO/KALYDECO

Start date ___________ ❑ Estimate

Dose: Age 2 to <6 <14kg Kaftrio 60/40/80 one sachet mane, Kalydeco 59.5 one sachet nocte ❑

Age 2 to <6 ≥14kg Kaftrio 75/50/100 one sachet mane, Kalydeco 75 one sachet nocte ❑

Age 6 to <12 <30kg Kaftrio 37.5/25/50 two tablets mane, Kalydeco 75 one tablet nocte ❑

Age 6 to <12 ≥30kg Kaftrio 75/50/100 two tablets mane, Kalydeco 150 one tablet nocte ❑

Age ≥12 Kaftrio 75/50/100 two tablets mane, Kalydeco 150 one tablet nocte ❑

Adjusted dose (e.g. liver disease or drug interaction): ____

	Pre-treatment	Annual
Sweat chloride (mmol/l)		
Sweat chloride (date)		
FEV_1 (date)		
FEV_1 (value)		
FEV_1 (% predicted)		

Are they still taking this drug: ❑ Yes ❑ No

ORKAMBI

Start date ___________ ❑ Estimate

Dose: 7 to <9kg Orkambi 75/94 one sachet BD ❑

9 to <14kg Orkambi 100/125 one sachet BD ❑

≥14kg Orkambi 150/188 one sachet BD100/125 1 sachet twice daily ❑

Age 6+ years Orkambi 100/125 two tablets BD ❑

Adjusted dose (e.g. liver disease/drug interaction): ____

	Pre-treatment	Annual
Sweat chloride (mmol/l)		
Sweat chloride (date)		
FEV_1 (date)		
FEV_1 (value)		
FEV_1 (% predicted)		

Are they still taking this drug: ❑ Yes ❑ No

IVACAFTOR

Start date ___________ ❑ Estimate

Dose: 1 to <3 mth, <3kg 13.4mg one sachet BD ❑

1 to <3 mth, ≥3kg 25mg one sachet BD ❑

≥6 mth, ≥5 to <7kg 25mg one sachet BD ❑

≥6 mth, ≥7 to <14kg 50mg one sachet BD ❑

≥6 mth, ≥14 to <25kg 75mg one sachet BD ❑

6+ years, 150 mg one tablet twice daily ❑

	Pre-treatment	Annual
Sweat chloride (mmol/l)		
Sweat chloride (date)		
FEV_1 (date)		
FEV_1 (value)		
FEV_1 (% predicted)		

Are they still taking this drug: ❑ Yes ❑ No

SYMKEVI Start date ___________ **Dose:** Symkevi 100/150 mane, Kaftrio 150mg nocte ❑

NURSE

CULTURE & MICROBIOLOGY

Number of sputum samples since last annual review ___________

Number of cough/throat/nasal samples since last annual review ___________

Number of bronchoscopy samples since last annual review ___________

Culture result:

❑ Positive culture sample ❑ No growth ❑ Normal flora ❑ Awaited

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Culture growth:

❑ Pseudomonas aeruginosa — Number of pseudomonas aeruginosa samples since last annual review: ___________

Pseudomonas mucoid status ❑ Mucoid ❑ Non mucoid ❑ Unknown

Pseudomonas drug resistance
- ❑ Pseudomonas multi drug resistant
- ❑ Pseudomonas other resistant pattern
- ❑ Pseudomonas fully sensitive
- ❑ Pseudomonas ciprofloxacin resistant
- ❑ Intermittent
- ❑ Chronic

		Number of samples
❑ Burkholderia cepacia complex	❑ Burkholderia cepacia	___________
	❑ Burkholderia cenocepacia	___________
	❑ Burkholderia multivorans	___________
	❑ Other Burkholderia cepacia species	___________

		Number of samples
❑ Staphylococcus aureus	❑ Chronic	___________
	❑ Intermittent	___________

		Number of samples
❑ Other cultures	❑ Alcaligenes (Achromobacter) xylosoxidans	___________
	❑ Pseudomonas species	___________
	❑ Escherichia coli	___________
	❑ Haemophilus influenza	___________
	❑ Klebsiella	___________
	❑ MRSA	___________
	❑ Pandoraea	___________
	❑ Stenotrophomonas (Xanthomonas) maltophilia	___________
	❑ Other (specify) ______________________	___________

		Number of samples
❑ Fungal	❑ Aspergillus fumigatus	___________
	❑ Scedosporium species	___________
	❑ Aspergillus species	___________
	❑ Candida	___________
	❑ Fungal Other (specify) ______________________	___________

❑ Viral ❑ SARS/COVID ❑ Influenza ❑ RSV

❑ Other (specify) ______________________

NTM

Has the patient been on any treatment for NTM pulmonary disease at any time since last annual review: ❑ Yes ❑ No
Has the patient had NTM positive samples since last annual review: ❑ Yes ❑ No

If no: Negative culture result: ❑ Negative culture sample ❑ No sample taken
❑ Contaminated culture sample ❑ Not known

If yes: Date of culture _______________

Culture type:
- ❑ Sputum
- ❑ Induced sputum
- ❑ Lung biopsy
- ❑ Broncho-alveolar lavage
- ❑ Not known

If treatment received, antibiotics prescribed:
- ❑ Amikacin IV ❑ Amikacin (nebs)
- ❑ Imipenem ❑ Clofazimine
- ❑ Tigecycline ❑ Minocycline
- ❑ Other ❑ Moxifloxacin
- ❑ Other

Species: ____

Has the patient been on oral corticosteroid since the last data set: ❑ Yes ❑ No ❑ Not known

COMPLICATIONS

CFRD Status: ❑ CFRD ❑ Steroid Induced Diabetes
❑ Impaired Glucose Tolerance ❑ Indeterminate
❑ No CFRD

Does patient have CF related diabetes (CFRD) diagnosis:
- ❑ No
- ❑ CFRD with fasting hyperglycaemia
- ❑ CFRD without fasting hyperglycaemia
- ❑ CFRD

CF related diabetes (CFRD) complications:
- ❑ None
- ❑ Diabetic retinopathy
- ❑ Diabetic microalbuminuria
- ❑ Other

Was patient prescribed treatment for CFRD: ❑ Yes ❑ No If yes:
- ❑ Dietary change
- ❑ Oral hypoglycaemic agents
- ❑ Intermittent insulin
- ❑ Chronic insulin

Has patient been newly diagnosis with a cancer since last annual review: ❑ Yes ❑ No
Septicaemia with positive blood cultures ❑ Yes ❑ No ❑ Unknown
Haemoptysis massive, severe or moderate ❑ Yes ❑ No Haemoptysis scanty (<=5 mls in 24 hours) ❑ Yes ❑ No

Any liver/gallbladder complications: ❑ Yes ❑ No
If yes:
- ❑ Gall bladder disease/stones
- ❑ Raised liver enzymes
- ❑ Liver disease

If yes to liver disease:

i. Is this CF related liver disease? ❑ Yes ❑ No
- ❑ Hepatic steatosis (fatty liver)
- ❑ Chronic liver disease with no cirrhosis (early fibrosis)
- ❑ Cirrhosis with portal hypertension
- ❑ Cirrhosis with no portal hypertension

Complication of portal hypertension:
- ❑ Bleeding from varices
- ❑ Oesophageal injection or banding
- ❑ Hypersplenism (WBC <3.0, Platelets <100)
- ❑ Ascites
- ❑ Hepatic encephalopathy

ii. Are there acute liver complications? ❑ Yes ❑ No

a. Acute liver failure (no underlying liver disease, ALT>3xULN, INR>2, not responsive to vitamin K): ❑ Yes ❑ No
b. Acute hepatitis (ALT >5xULN, and duration of illness <6 mths) ❑ Yes ❑ No

i. If acute hepatitis: Infectious cause? ❑ Yes ❑ No
Drug induced ❑ Yes ❑ No
Suspected drug ___________ ❑ Unknown

Were there any gut complications: ❑ Yes ❑ No

If yes:
- ❑ DIOS (Distal Intestinal Obstruction Syndrome)
- ❑ Fibrosis colonopathy/colonic stricture
- ❑ Intestinal obstruction
- ❑ Gastro-oesophageal reflux disease
- ❑ Gastrointestinal non-varices as source
- ❑ Pancreatitis
- ❑ Peptic ulcer
- ❑ Rectal prolapse

Were there any kidney/renal complications: ❑ Yes ❑ No If yes:
- ❑ Hypertension
- ❑ Acute kidney injury requiring dialysis
- ❑ Kidney stones
- ❑ Chronic renal failure

ABPA ❑ Yes ❑ No

Highest IgE result since last AA ______________ Date of result ________________________

ABPA Treated? ❑ Yes ❑ No

❑ Steroid
❑ Azole Antifungals
❑ Nebulised Amphoteracin
❑ Other ____

Are there any other complications: ❑ Yes ❑ No

If yes:

❑ Arthritis
❑ Arthropathy
❑ Allergic Broncho Pulmonary Aspergillosis
❑ Asthma
❑ Bone fracture
❑ Depression
❑ Hearing loss
❑ Nasal polyps
❑ Osteopenia
❑ Osteoporosis
❑ Paediatric intensive care unit
❑ Pneumothorax requiring chest drain
❑ Port inserted or replaced since last annual review
❑ Sinus disease
❑ Other (specify) ___________________

DIETITIAN

GROWTH & NUTRITION

Nutritional assessment carried out this encounter? ❑ Yes ❑ No
Seen by specialist CF dietitian: ❑ Yes ❑ No
Assessed for oral intake: ❑ Yes ❑ No

Supplemental feeding:

❑ None
❑ Oral
❑ Nasogastric
❑ Gastrostomy
❑ Jejunal tube
❑ Parental
❑ Yes but method unknown
❑ Not known

Dose of lipase: ______________ (IU/kg/per day) ❑ Not known ❑ Not applicable

Does the patient take pancreatic enzyme supplements: ❑ Yes ❑ No ❑ Not known
Has the patient been on oestrogen/testosterone: ❑ Yes ❑ No ❑ Not known

Dietetic Summary

Dietetic summary to be dictated

PHYSIOTHERAPY

Airway Clearance
(tick all that apply)

Primary	*Secondary*
❑ Active Cycle Breathing Technique	❑ Active Cycle Breathing Technique
❑ Assisted autogenic drainage	❑ Assisted autogenic drainage
❑ Autogenic drainage	❑ Autogenic drainage
❑ Exercise	❑ Exercise
❑ Forced expiration	❑ Forced expiration
❑ High pressure PEP	❑ High pressure PEP
❑ Oscillating PEP	❑ Oscillating PEP
❑ PEP	❑ PEP
❑ Postural drainage	❑ Postural drainage
❑ Vest	❑ Vest
❑ None	❑ None
❑ Other	❑ Other

Used non-invasive ventilation: ❑ Yes ❑ No ❑ Not known
Has an exercise test been performed: ❑ Yes ❑ No ❑ Not known

If yes: ❑ Submaximal ❑ Walk test ❑ Other (specify) ___________________
❑ Shuttle test ❑ Step test

Urinary incontinence: ❑ Yes ❑ No ❑ Not known
Faecal incontinence: ❑ Yes ❑ No ❑ Not known
Postural abnormality: ❑ Yes ❑ No ❑ Not known

Physiotherapy Summary

Physiotherapy summary to be dictated

LIFESTYLE

Social History

Psychosocial Issues

Does the patient smoke cigarettes or other forms of tobacco:

- ❑ Declined to answer
- ❑ No
- ❑ Not asked
- ❑ Yes, occasionally
- ❑ Yes, regularly <1 pack per day
- ❑ Yes, regularly 1 pack per day or more
- ❑ Yes, amount unknown
- ❑ Not known

Is the patient regular exposed to second hand smoke or vaping: ❑ Yes ❑ No ❑ Not known
Does the patient use e-cigarettes: ❑ Yes ❑ No ❑ Not known

Allowances

DLA: ❑ High ❑ Middle ❑ Low
Mobility: ❑ High ❑ Low
Carer's Allowance: ❑ Yes ❑ No

Nursery/School

Name of school ______________________________
Head teacher ______________________________
Class teacher ______________________________
Attendance ______________________________

Nursing Summary

Nursing summary to be dictated

Consultant Summary & Recommendations

Consultant Summary & Recommendations

Psychology Review

QOL Questionnaire Completed ❑ Yes ❑ No

❑ 6 – 11 ❑ 12 – 13 ❑ Teens/Adults (14 and over) ❑ Care giver