CHD Clinical Practice Algorithm: First-Year Post Fontan[1-5]

Inclusion criteria:
- <12 years of age
- Status post Fontan
- Clinically stable

Standard surgical follow-up:
- Post-op ECG, TTE, and CXR done prior to discharge

Clinic follow-up within 2 weeks to 1 month:
- Physical examination
- sO_2
- TTE ± ECG
- CXR

Identify:
- Pleural effusions
- Change in ventricular dysfunction/AVVR
- Cyanosis
- Arrhythmia

Yes → Further evaluation and clinical management based on presentation[a]

No →

Follow-up every 3-6 months with:
- Physical examination
- sO_2
- ± ECG
- ± TTE

until 1 year post Fontan

Therapies[b]

All Patients

Antiplatelet/anticoagulation medications
- **No** previous TE: aspirin, warfarin, or DOAC
- **Yes** previous TE: warfarin or DOAC

Lifestyle counseling
- Encourage and counsel on exercise and physical activity

Education on Fontan-related complications

Consultation
- Proactive social work/psychology consultation

Select Patients

CV medications
- ACEI/ARB
- Aldosterone antagonist
- PDE5i or ERA

Consultation
- Proactive HF/advanced therapies consultation

Evaluation/surveillance:
Go to page 2

[a]See text for encountered issues/ organ systems

[b]Therapies listed are intended to represent the most common therapies/approaches relevant to most patients, but do not represent a comprehensive set of all Fontan-related therapies for clinically stable individuals

Abbreviations:
ACEI = angiotensin converting enzyme inhibitor; ARB = angiotensin II receptor blocker; AVVR = atrioventricular valve regurgitation; CHD = congenital heart disease; CV = cardiovascular; CXR = chest X-ray; DOAC = direct oral anticoagulant; ECG = electrocardiogram; ERA = endothelin-receptor antagonist; HF = heart failure; PDE5i = phosphodiesterase-5 inhibitor; post-op = postoperative; sO_2 = oxygen saturation; TE = thromboembolism; TTE = transthoracic echocardiogram.

CHD Clinical Practice Algorithm: Fontan <12 Years of Age[1-3, 6]

Inclusion criteria:
- <12 years of age
- Status post Fontan
- Clinically stable

Evaluation/surveillance:

Clinic follow-up every 6-12 months
- Clinic visit
- sO_2
- ECG
- TTE

Every 2-3 years
- Ambulatory arrhythmia monitor
- CPET starting at ~9-10 years of age or when developmentally appropriate

Every 3-4 years
- Serum BNP or NT-proBNP level
- Basic labs: CMP, serum GGT, CBC, serum cystatin C, vitamin D levels
- cMRI (as clinically indicated and when able without sedation, ~11 years of age)
- Abdominal US with Doppler, US elastography (standard choice)
- Abdominal MRI elastography (in those who have previously had MRI for trending)
- Referral for neurodevelopmental/ cognitive testing

Consider referral to specialized multidisciplinary Fontan clinic if available

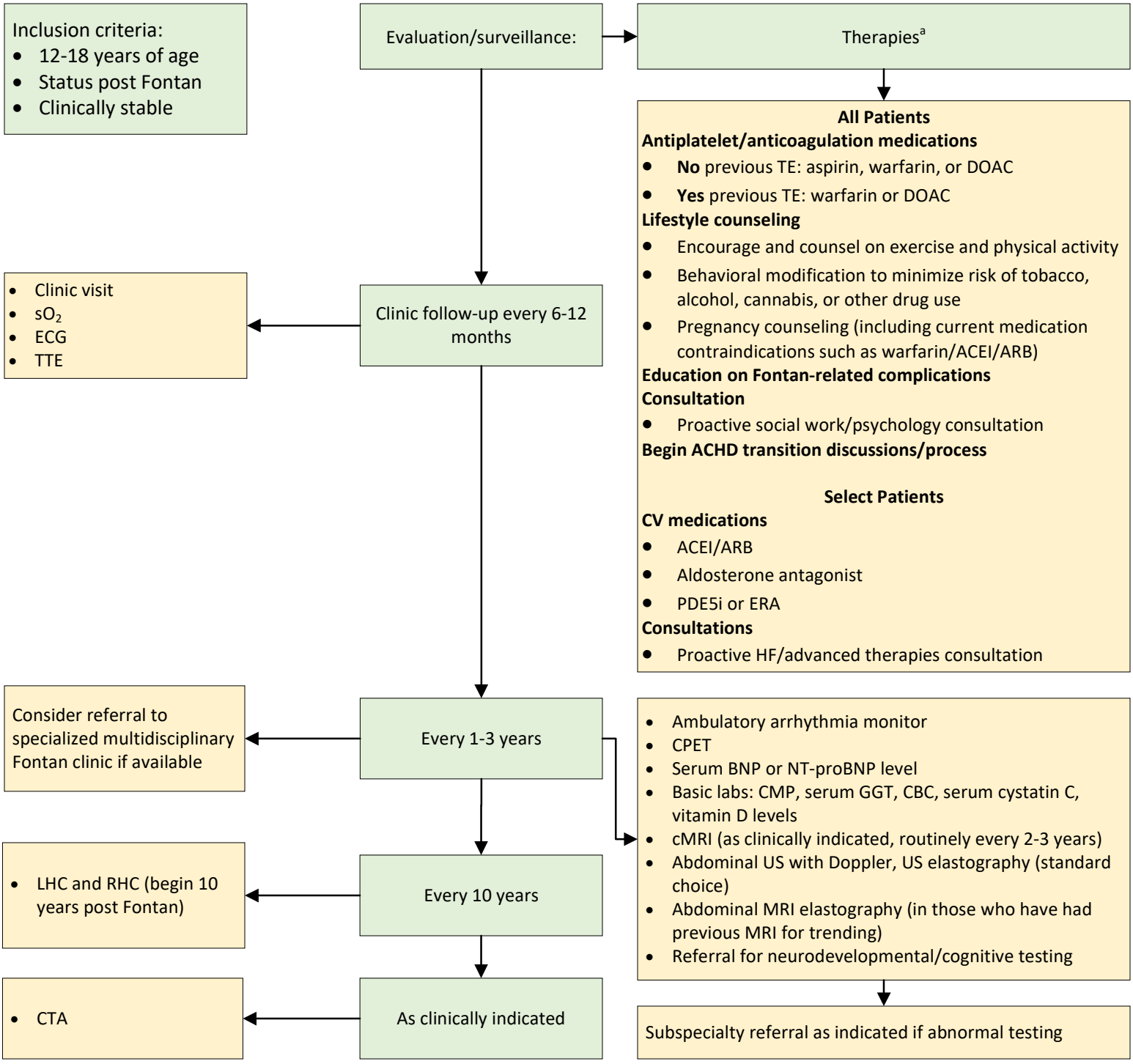
Subspecialty referral as indicated if remarkable testing results

As clinically indicated
- CTA
- LHC and RHC

Note: Refer to the therapies box on page 1

Abbreviations:
BNP = B-type natriuretic peptide; CBC = complete blood count; CHD = congenital heart disease; CMP = comprehensive metabolic profile; cMRI = cardiac magnetic resonance; CPET = cardiopulmonary exercise testing; CTA = computed tomography angiography; ECG = electrocardiogram; GGT = gamma-glutamyl transferase; labs = laboratory studies; LHC = left heart catheterization; MRI = magnetic resonance; NT-proBNP = N-terminal pro–B-type natriuretic peptide; RHC = right heart catheterization; sO_2 = oxygen saturation; TTE = transthoracic echocardiogram; US = ultrasonography.

CHD Clinical Practice Algorithm: Fontan 12-18 Years of Age[1-6]



[a]Therapies listed are intended to represent the most common therapies/approaches relevant to most patients, not a comprehensive list of all Fontan-related therapies for clinically stable individuals

Abbreviations:
ACEI = angiotensin converting enzyme inhibitor; ACHD = adult congenital heart disease; ARB = angiotensin II receptor blocker; BNP = B-type natriuretic peptide; CBC = complete blood count; CHD = congenital heart disease; CMP = comprehensive metabolic profile; cMRI = cardiac magnetic resonance; CPET = cardiopulmonary exercise testing; CTA = computed tomography angiography; DOAC = direct oral anticoagulant; ECG = electrocardiogram; ERA = endothelin-receptor antagonist; HF = heart failure; GGT = gamma-glutamyl transferase; labs = laboratory studies; LHC = left heart catheterization; MRI = magnetic resonance; NT-proBNP = N-terminal pro–B-type natriuretic peptide; PDE5i = phosphodiesterase-5 inhibitor; RHC = right heart catheterization; sO_2 = oxygen saturation; TE = thromboembolism; TTE = transthoracic echocardiogram; US = ultrasonography.

CHD Clinical Practice Algorithm: Fontan ≥18 Years of Age[2-5,7-10]

Inclusion criteria:
- Age ≥18 years
- Status post Fontan operation without subsequent takedown
- Clinically stable (NYHA class I-II HF, no PLE/PB, no recent acute events/hospitalizations (e.g., over previous 12 months)

First visit at ACHD clinic:
- Communicate with pediatric cardiologist
- Obtain all operation reports, cath reports, EP reports, liver imaging/pathology and most recent TTE/cath images
- Discuss differences between pediatric/adult care in the local context

Clinic follow-up every 6-12 months
- Clinic visit
- sO_2[b]
- ECG
- TTE[c]
- BNP/NT-proBNP[c]
- CBC
- CMP
- PT/INR[c]
- Cystatin C[c]
- Alpha-fetoprotein

Every 1-2 years
- Ambulatory arrhythmia monitoring
- CPET[d]

Every 3 years
- cMRI
- Abdominal MRI with elastography[e]

Every 10 years
- LHC and RHC[f]

At least once
- Hepatology referral
- Labs for hepatitis A/B/C
- PFT

Therapies[a]

All Patients

Antiplatelet/anticoagulation medications
- **No** previous TE: aspirin, warfarin, or DOAC
- **Yes** previous TE: warfarin or DOAC

Immunizations
- In particular, immunization for hepatitis A/B

Lifestyle counseling
- Encourage/counsel on exercise and physical activity
- Behavioral modification (e.g., to ameliorate risk of tobacco, alcohol, and other harmful substance use)
- Pregnancy counseling (including current medication contraindications such as warfarin/ACEI/ARB/ARNI)
- Mental health check-in/discussion

Education on Fontan-related complications

Consultation
- Proactive social work/psychology consultation

Select Patients

CV medications
- ACEI/ARB/ARNI
- Aldosterone antagonist
- SGLT2i
- BB
- PDE5i or ERA

Consultation
- Proactive HF/advanced therapies consultation

[a]Therapies listed are intended to represent the most common therapies/approaches relevant to most patients, not a comprehensive list of all Fontan-related therapies for clinically stable individuals

[b]Consider cardiac cath if unexplained/worsening hypoxemia

[c]Timing of TTE may vary. In the absence of clinical change, TTE does not need to be repeated >1/year. If on warfarin, INR will be more frequent. BNP/NT-proBNP and cystatin C assay may be performed less frequently if clinically stable. These labs are not comprehensive and others may be appropriate (e.g., 25-OH vitamin D, parathyroid hormone, IgG).

[d]Consider more often if medication change or clinical concerns

[e]If contraindication to MRI, may consider US alternating with CT

[f]Consider earlier than every 10 years if change in clinical status

Abbreviations:
25-OH = 25-hydroxy; ACEI = angiotensin converting enzyme inhibitor; ACHD = adult congenital heart disease; ARB = angiotensin II receptor blocker; ARNI = angiotensin receptor–neprilysin inhibitor; BB = beta-blocker; BNP = B-type natriuretic peptide; cath = catheterization; CBC = complete blood count; CHD = congenital heart disease; CMP = comprehensive metabolic profile; cMRI = cardiac magnetic resonance; CPET = cardiopulmonary exercise testing; CT = computed tomography; CV = cardiovascular; DOAC = direct oral anticoagulant; ECG = electrocardiogram; EP = electrophysiology; ERA = endothelin-receptor antagonist; HF = heart failure; IgG = immunoglobulin G; INR = international normalized ratio; labs = laboratory studies; LHC = left heart catheterization; MRI = magnetic resonance; NT-proBNP = N-terminal pro–B-type natriuretic peptide; NYHA = New York Heart Association; PB = plastic bronchitis; PDE5i = phosphodiesterase-5 inhibitor; PFT = pulmonary function testing; PLE = protein losing enteropathy; PT = prothrombin time; RHC = right heart catheterization; SGLT2i = sodium-glucose cotransporter-2 inhibitor; sO_2 = oxygen saturation; TE = thromboembolism; TTE = transthoracic echocardiogram; US = ultrasonography.

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