

MDAnderson Li-Fraumeni Syndrome Screening - Pediatric

Page 1 of 4

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Note: Screening is only intended for asymptomatic individuals. Individuals undergoing Li-Fraumeni Syndrome screening should have no co-morbidities that would limit the diagnostic evaluation or treatment of any identified problem. The screening technique should be performed with a consistent technique and process.

PRESENTATION ASSESSMENT RECOMMENDATION Initiate cancer screening³: Personal and/or family history • Refer to appropriate oncology • Referral to Pediatric LFS Screening Clinic indicative of Li-Fraumeni team based on finding for assessment/evaluation Syndrome (LFS) or family • Continue cancer screening for o Complete physical exam Yes member diagnosed with LFS o Symptom assessment; review findings Positive other body sites (see Page 2) through genetic testing o Provide patient education⁴ clinical symptom or finding upon o Psychologic evaluation and counseling • Lifestyle risk assessment⁵ screening? No Yes o Review personal and family history to Continue cancer screening Referral to Genetic identify additional at-risk relatives and (see Page 2) testing indicates **Pediatric** provide recommendations for counseling/ germline TP53 Genetics for testing mutation²? genetic testing No Cancer screening may be tailored based on personal and family history, per discussion with healthcare team including Genetic Counselor

Offspring of a parent with LFS or personal and/or family history of LFS-associated cancers including adrenocortical carcinomas, breast cancer, central nervous system tumors, osteosarcomas, and soft-tissue sarcomas. Additional LFS-associated cancers include leukemia, lymphoma, gastrointestinal cancers, cancers of head and neck, kidney, larynx, lung, skin (e.g., melanoma), ovary, pancreas, prostate, testis, and thyroid. See CRIT-7 LFS testing criteria within the NCCN guidelines.

² If genetic test results do not clearly indicate whether the TP53 mutation is germline, mosaic or somatic in nature, refer to Li-Fraumeni Syndrome Screening - Adult algorithm for process of confirmatory testing

³ See Screening Guidelines on Page 2

⁴ Refer to Patient Education: Li-Fraumeni Syndrome (LFS) Screening Program - Pediatric Screening Program

⁵ See Physical Activity, Nutrition, Obesity Screening and Management, and Tobacco Cessation Treatment algorithms; ongoing reassessment of lifestyle risks should be a part of routine clinical practice



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Page 2 of 4

Cancer Center

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Li-Fraumeni Syndrome Screening Program – Pediatric Screening Guidelines

Cancer	Age			Frequency
	< 1 Year Old	1-9 Years Old	10-20 Years Old	
General	Physical exam/targeted review of systems • Neurological exam	Physical exam/targeted review of systems • Neurological exam	Physical exam/targeted review of systems • Neurological exam • Skin	Every 6 to 12 months
Adrenocortical Tumor (ACT) and Others	 Education of signs and symptoms (virilization, Cushing's Syndrome, hypertension) Testosterone, DHEA-S, ACTH 	 Education of signs and symptoms (virilization, Cushing's Syndrome, hypertension) Testosterone, DHEA-S, ACTH 	 Education of signs and symptoms (virilization, Cushing's Syndrome, hypertension) Testosterone, DHEA-S, ACTH 	Every 6 months (until 10 years old)Annually (10-20 years old)
	Ultrasound of abdomen and pelvis	Ultrasound of abdomen and pelvis	MRI whole body	Every 6 months for ultrasoundAnnually for MRI whole body
Brain	 Education of signs and symptoms (vomiting, headaches, vision changes) MRI¹ brain 	 Education of signs and symptoms (vomiting, headaches, vision changes) MRI¹ brain 	 Education of signs and symptoms (vomiting, headaches, vision changes) MRI¹ brain 	Annually
Sarcoma (begin at 2-3 years of age based on family history/clinical judgement)	N/A	MRI whole body	MRI whole body	Annually
Leukemia/ Lymphoma	 Education of signs and symptoms (anemia, pallor, fatigue, bruising, petechiae) CBC with differential, lactate dehydrogenase 	 Education of signs and symptoms (anemia, pallor, fatigue, bruising, petechiae) CBC with differential, lactate dehydrogenase 	 Education of signs and symptoms (anemia, pallor, fatigue, bruising, petechiae) CBC with differential, lactate dehydrogenase 	• Every 6 months (until 10 years old) • Annually (10-20 years old)
Melanoma	N/A	N/A	Refer to Dermatology service as necessary ²	Annually

DHEA-S = dehydroepiandrosterone-sulfate ACTH = adrenocorticotropic hormone

Department of Clinical Effectiveness V5

¹ First MRI with contrast using gadopiclenol (Vueway[®]); thereafter without contrast if previous MRI is negative with no new abnormality; an exception is when the MRI of the brain is performed in conjunction with the MRI of the whole body: if so, the MRI of the brain should be performed after the MRI of the whole body and take advantage of the gadopiclenol (Vueway[®]) injected during the MRI of the whole body.

² Refer to Dermatology if there is a concern about a skin lesion or if there is a family member diagnosed with cutaneous melanoma at a young age

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MD Anderson Li-Fraumeni Syndrome Screening - Pediatric

Page 3 of 4

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SUGGESTED READINGS

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Making Cancer History®

MD Anderson Li-Fraumeni Syndrome Screening - Pediatric

Page 4 of 4

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DEVELOPMENT CREDITS

This screening algorithm is based on majority expert opinion of the Pediatric Li-Fraumeni Syndrome workgroup at the University of Texas MD Anderson Cancer Center. It was developed using a multidisciplinary approach that included input from the following:

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