Li-Fraumeni Syndrome Screening - Pediatric

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**PRESENTATION**

Personal and/or family history indicative of Li-Fraumeni syndrome

- Referral to Genetics for genetic testing

**ASSESSMENT**

- Genetic testing indicates germline p53 mutation?
  - Yes
    - Initiate cancer screening:
      - Referral to Pediatric LEAD clinic for assessment/evaluation
      - Complete physical exam
      - Symptom assessment; review findings
      - Provide patient education
      - Psychologic evaluation and counseling
      - Lifestyle risk assessment

  - No
    - Cancer screening may be tailored based on personal and family history, per discussion with Genetic Counselor

- Positive clinical symptom or finding upon screening?
  - Yes
    - Refer to appropriate oncology team based on finding
    - Continue cancer screening for other body sites
  - No
    - Continue cancer screening

**RECOMMENDATION**

LEAD clinic = Li-Fraumeni Syndrome Education and Early Detection clinic

1 See Screening Guidelines on page 2
2 Patient Education - Li-Fraumeni Syndrome Education and Early Detection (LEAD) Pediatric Screening Program
3 See Physical Activity, Nutrition, and Tobacco Cessation algorithms; ongoing reassessment of lifestyle risks should be a part of routine clinical practice

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## Li-Fraumeni Syndrome Education and Early Detection (LEAD) – Pediatric Screening Guidelines

<table>
<thead>
<tr>
<th>Cancer</th>
<th>0-1 Year</th>
<th>1-10 Years</th>
<th>10-20 Years</th>
<th>Frequency</th>
</tr>
</thead>
</table>
| 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  
Thyroid  
Skin | Every 6 months |
| Adrenocortical Tumor (ACT) and Others | Education of signs and symptoms (virilization, Cushing’s syndrome, hypertension)  
Testosterone, DHEAS, HCG, AFP, ACTH, urinalysis | Education of signs and symptoms (virilization, Cushing’s syndrome, hypertension)  
Testosterone, DHEAS, HCG, AFP, ACTH, urinalysis | Education of signs and symptoms (virilization, Cushing’s syndrome, hypertension)  
Testosterone, DHEAS, HCG, AFP, ACTH, urinalysis | Every 6 months (until 10 years old)  
Annually (10 – 20 years old) |
| Ultrasound of abdomen and pelvis | Ultrasound of abdomen and pelvis | Whole body MRI¹ | Every 6 months for ultrasound  
Annually for whole body MRI¹ |
| Brain | Education of signs and symptoms (vomiting, headaches, vision changes)  
Brain MRI¹,² | Education of signs and symptoms (vomiting, headaches, vision changes)  
Brain MRI¹,² | Education of signs and symptoms (vomiting, headaches, vision changes)  
Brain MRI¹,² | Annually |
| Sarcoma (begin at 2-3 years – based on family history/clinical judgement) | N/A | Whole body MRI¹ | Whole body MRI¹ |
| Leukemia/ Lymphoma | Education of signs and symptoms (anemia, pallor, fatigue, bruising, petechiae)  
CBC, erythrocyte sedimentation rate, lactate dehydrogenase | Education of signs and symptoms (anemia, pallor, fatigue, bruising, petechiae)  
CBC, erythrocyte sedimentation rate, lactate dehydrogenase | Education of signs and symptoms (anemia, pallor, fatigue, bruising, petechiae)  
CBC, erythrocyte sedimentation rate, lactate dehydrogenase | Every six months (until 10 years)  
Annually (10 – 20 years old) |
| Melanoma | N/A | N/A | Refer to Dermatology service as necessary | Annually |

1 BUN and creatinine prior to any MRI  
2 First MRI with contrast; thereafter without contrast if previous MRI normal and no new abnormality

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SUGGESTED READINGS


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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