Inclusion Criteria
- Patients with VSD of any size with or without minor additional defects (PPS, PFO, small PDA)
- Younger than 1 year old at diagnosis
- Greater than 38 weeks corrected gestational age

Exclusion Criteria
- 1 year old or older at diagnosis
- Genetic syndromes affecting surgical timing (i.e. Trisomy 21)
- Hemodynamically significant heart disease (Coarctation, LVOT obstruction, AV septal defect-complete or transitional)
**VSD v.2.0: Unrestrictive VSD**

**Executive Summary**

**Test Your Knowledge**

**Explanation of Evidence Ratings**

**Summary of Version Changes**

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**Initiate Therapy**
- Furosemide 1 mg/kg per dose two times a day
- Increase calories to 24-26 kcal/ounce

**Follow in 2-4 weeks**

**Acceptable Symptoms:**
(+/− Tachypnea with adequate weight gain defined as 15-30g/day)

**YES**

**YES**

**Follow at 2-4 week intervals until age 4 months and repeat single ECHO**

**NO**

**NO**

**YES**

**Escalate Therapy**
- Furosemide 1mg/kg per dose three times a day
- Consider adding spironolactone or captopril
- Follow up lytes, BUN,Cr in 1 week
- Repeat ECHO

**YES**

**Unrestrictive VSD?**

**YES**

**Present for Surgery**

**NO**

**Follow every 2-3 months for acceptable symptoms**

---

**Congestive Heart Failure (Tachypnea +/- Fail to thrive)**

**Follow at 4 week intervals**

**YES**

**NO**

**Follow at 4 week intervals until age 4 months and repeat single ECHO**

**NO**

**Unrestrictive VSD?**

**YES**

**See back at 12 months of age or as clinically indicated**

**NO**

---

**Delay VSD surgery for 4-6 weeks after a positive viral PCR/culture or for any other clinically significant illness**

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VSD v.2.0: Restrictive VSD

**Executive Summary**

- Restrictive VSD: VSD Velocity >3 m/s AND RV pressure < 50% systemic

**Test Your Knowledge**

- Acceptable Symptoms: +/- Tachypnea with adequate weight gain defined as 15-30g/day

**Citation Information**

- Indications for surgery: symptoms despite two times a day furosemide, NG feeds, hospitalization for congestive heart failure, inadequate weight gain, persistent unrestrictive VSD at 4 months of age
  - If symptoms improve, repeat echocardiogram at 6 months and consider presenting for surgery if still large VSD or weaning of medication is unlikely

For questions concerning this pathway, contact: VSD@seattlechildrens.org

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VSD v.2.0: Post-op VSD Closure Discharge

Executive Summary

Test Your Knowledge

Citation Information

Explanation of Evidence Ratings

Summary of Version Changes

Inclusion Criteria
- Patients with surgical closure of VSD AND younger than 1 year of age

Exclusion Criteria
- Hospital length of stay > 7 days
- 1 year of age or Older

Discharge Criteria
- Tolerating all prescribed medications
- Tolerating feeding regimen
- ECHO and ECG, expected post operative changes without residual defect or unexpected abnormality
- Stable labs
- All equipment, medication and formula, instructions are provided to the parents/family

Discharge Medications
Diuretics: Furosemide 1 mg/kg per dose 2 times per day
Bowel regimen:
- Simethicone 20mg PO 4 times/day PRN flatulence
- Glycerin 0.5 suppository rectally once/day PRN constipation
Pain regimen:
<6 months of age:
- Acetaminophen 12.5mg/kg PO every 4 hours PRN pain x 5 days
- Morphine 0.1mg/kg PO every 4 hours PRN pain x 5 days
>6 months of age:
- Acetaminophen 12.5mg/kg PO every 4 hours PRN pain x 5 days
- Oxycodone 0.1 mg/kg PO every 4 hours PRN pain x 5 days

Discharge Appointments
- Cardiac Surgery or Cardiology, 7-10 days; chest x-ray and ECG at this appointment
- Wean furosemide to once/day
- Primary Care Provider, 1-2 weeks
- Cardiology, 2-4 weeks, stop Furosemide; ECHO at this appointment

Off Pathway Indications:
- Mediastinitis
- Post pericardiotomy syndrome
- Large residual VSD
There are no clear guidelines on the medical management of the infant with a symptomatic VSD.

Initial medical treatment will consist of:
- Furosemide 1 mg/kg per dose given two times per day
- Increase calories to 24-26 kcal/ounce (formula or breast milk)

Escalation of medical therapy will include:
- Furosemide 1 mg/kg per dose given three times per day
- Consideration of the following:
  - Spironolactone- Start at 0.5 mg/kg/dose 3 times per day
  - Captopril- Start at 0.1mg/kg/dose, titrate dose upward to maximum of 2mg/kg/dose
- Monitoring of electrolytes and renal function

[LOE: Expert Opinion]
Ventricular Septal Defect: Surgical Management

- Approximately 15-20% of the 10,000 infants diagnosed annually in the US with isolated VSD will require surgery
- No clear guidelines on the optimal timing for surgical closure of the VSD
- VSD surgical closure is most appropriate in patients > 3 kg with unacceptable symptoms of CHF or persistent unrestrictive physiology

[LOE: Expert Opinion]

Ventricular Septal Defect: Surgical Management (Cont'd)

- Indications for surgery:
  - Symptoms despite two times a day furosemide
  - Hospitalization for congestive heart failure
  - Inadequate weight gain despite caloric intake of >120 kcal/kg/d
  - Requirement for NG Feeds
  - Persistent unrestrictive VSD at 4 months of age

[LOE: Expert Opinion]

- Reasons to delay surgery:
  - Delay VSD surgery for 4-6 weeks after a positive viral PCR/culture or for any other clinically significant respiratory illness.

[LOE: Expert Opinion]
Discharge Criteria: Following Surgical Closure of VSD

Discharge Criteria
- Tolerating all prescribed medications
- Tolerating feeding regimen
- ECHO and ECG, expected post operative changes without residual defect or unexpected abnormality
- Stable labs
- All equipment, medication and formula, instructions are provided to the parents/family

[LOE: Expert Opinion]
Clinical Follow-up After Surgical VSD Closure

- There are no clear guidelines on the optimal clinical follow-up after discharge from VSD closure:
  - Patients will be discharged on furosemide 2 times per day.
  - Follow-up discharge appointments will include:
    - 7-10 days with cardiac surgery or cardiology; chest x-ray and ECG at this appointment. Plan to wean furosemide to daily
    - 1-2 weeks with primary care provider
    - 2-4 weeks with cardiology; ECHO at this appointment. Plan to discontinue furosemide

[LOE: Expert Opinion]
Objective
To standardize the care of infants with ventricular septal defect

Through adherence to the VSD clinical pathway, outpatients experiencing congestive heart failure and growth failure will have a decrease in the time spent with poor growth by 30%

Recommendations

1. VSD Surgical closure is most appropriate in patients > 3 kg with unacceptable symptoms of CHF or persistent unrestrictive physiology.

2. Perioperative VSD patients in congestive heart failure and growth failure will receive 24-26 kcal/ounce of formula/breast milk.

3. Initiate furosemide 1mg/kg/dose two times a day for VSD patients with congestive heart failure.

4. Increase furosemide to 1mg/kg/doses three times a day for patients with unimproved congestive heart failure.

5. Indications for VSD surgery include:
   • symptoms despite BID furosemide
   • NG feeds
   • hospitalization for CHF
   • inadequate weight gain
   • persistent unrestrictive VSD at 4 months of age

6. Delay VSD surgery for 4-6 weeks after a positive viral PCR/culture or for any other clinically significant illness.

7. Discharge criteria for post surgical VSD closure patients include:
   • Tolerating all prescribed medications
   • Tolerating feeding regimen
   • ECHO & ECG, expected post-operative changes without residual defect or unexpected abnormality
   • Stable labs
   • All equipment, medication and formula instructions provided to parents/family

8. Discharge post surgical VSD closure patients on furosemide 1mg/kg/dose two times per day.

9. Follow-up discharge appointments for post surgical VSD patients include:
   • 7-10 days with cardiac surgery or cardiology; chest x-ray and ECG at this appointment
Executive Summary

- 1-2 weeks with primary care provider
- 2-4 weeks with cardiology; ECHO at this appointment

Rationale
- Safety will be improved by standardization of medication dosages and by continuous monitoring for serious safety events for any patients receiving pathway care.
- Quality of care will improve by reducing variability with standardization, the use of the best available evidence and consensus.
- Delivery of care is anticipated to be more efficient with the use of multi-phased powerplans.
- Engagement is grounded in the fact that the pathway has been developed, reviewed, and vetted by all members of the medical team.
- Patient/Family Satisfaction will be addressed by implementing clinical standard work that will assure the highest quality of care.
- Costs will be monitored and presumed to be more predictable with standardization.

Evidence
A literature search was conducted by our librarian services in attempt to answer these clinical questions with the highest level of evidence. These references were further reviewed and their applicability to these questions summarized and documented to inform recommendations. Please see the Evidence and Recommendations document for specific details.

Implementation Items
- The development of a clinical algorithm and powerplan to promote the standardized care of patients with ventricular septal defect
- The development of training modules for staff education.
- The development of standardized patient and family education materials.

Metrics Plan
1. Count and length of stay for patients admitted to the hospital for symptomatic congestive heart failure (normalized to reflect the # of discharges per pt before/after implementation of clinical pathway)
2. Median Length of Stay following surgical closure before and after implementation of clinical pathway
3. Charges before/after implementation of clinical pathway including inpatient and outpatient charges
4. Number of echocardiograms performed per patient; including comparison before and after implementation of clinical pathway
5. Median age and weight at surgery; including comparison before and after implementation of clinical pathway
6. Number of visits per patient (overall and comparison of those requiring surgery with those who didn’t)

7. Percent of patient receiving furosemide 1mg/kg/dose two times/day at the time of hospital discharge following surgical closure of VSD

*Other process metrics are currently in development and will be tracked on the quarterly progress report. For more information please email VSD@seattlechildrens.org.*

**PDCA Plan**
The CSW owner and committee will follow metrics, continue to review medical literature, and make alterations to the pathway as needed.

**Revision History**
Date Approved: May 2014
Next Review Date: May 2017
Approved by the CSW VSD Pathway Team in May 2014

**CSW VSD Team:**

**Cardiology Physician Owner:** Jack Salerno, MD  
Heart Center Nurse Practitioner: Jennie Choe, ARNP  
Cardiology Physician: James Christiansen, MD  
Cardiology Physician: Matt Files, MD  
CICU Physician: Rob Mazor, MD  
Cardiology Physician: Sue Stephenson, MD  
Pharmacist: Rapheus Villanueva, PharmD  
Cardiology Clinic Nurse: Kendra Waldburger, RN

**Clinical Effectiveness Team:**

Consultant: Jeff Foti, MD  
Project Leader: Kate Drummond, MS, MPA  
KM Analyst: Nate Deam, MHA  
CIS Informatician: Mike Leu, MD, MS, MPA  
CIS Analyst: Heather Marshall  
CNS: Ashley Wagner, MN, RN, PCNS-BC, CPN  
Librarian: Susan Klawansky, MLS  
Program Coordinator: Ashlea Tade
Title: Ventricular Septal Defect (VSD) Pathway

Authors:
- Seattle Children’s Hospital
- Jack Salerno
- Matthew Files
- Jennie Choe
- Kate Drummond
- Jeff Foti
- Caren Goldenberg
- Robert Mazor
- Kendra Waldburger

Date: May 14, 2014

Retrieval Website: http://www.seattlechildrens.org/pdf/vsd-pathway.pdf

Example:
Completion qualifies you for 1 hour of Category II CME credit. If you are taking this self-assessment as a part of required departmental training at Seattle Children’s Hospital, you MUST logon to Learning Center.

**True/False:**

1) Surgery should be delayed 6 weeks after significant viral respiratory illness (T or F)

2) For the purpose of this clinical pathway, an unrestricted VSD is defined as a VSD velocity <3 m/s OR RV Pressure >50% systemic (T or F)

3) Initial medical treatment will consist of Furosemide 1mg/kg per dose given 2 times per day (T or F)

4) Patients with hemodynamically significant heart disease (LV outflow obstruction) should be excluded from the pathway (T or F)

5) Patient's of any age be enrolled in the pathway (T or F)

**Multiple Choice:**

6) Which of the following is **not** an indication for VSD surgery:

   A) Symptoms despite two times a day Furosemide
   
   B) Inadequate weight gain despite caloric intake of >120 kcal/kg/d
   
   C) Persistent, unrestricted VSD at 4 months of age
   
   D) Left atrial dilation on echocardiography
Answer Key

True/False:

1) True
2) False
3) True
4) True
5) False

Multiple Choice:

6) D
We used the GRADE method of rating evidence quality. Evidence is first assessed as to whether it is from randomized trial, or observational studies. The rating is then adjusted in the following manner:

Quality ratings are **downgraded** if studies:
- Have serious limitations
- Have inconsistent results
- If evidence does not directly address clinical questions
- If estimates are imprecise OR
- If it is felt that there is substantial publication bias

Quality ratings can be **upgraded** if it is felt that:
- The effect size is large
- If studies are designed in a way that confounding would likely underreport the magnitude of the effect OR
- If a dose-response gradient is evident

**Quality of Evidence:**
- ★★★★★ High quality
- ★★★★ Moderate quality
- ★★★ Low quality
- ★★ Very low quality
- ★ Expert Opinion (E)

Summary of Version Changes

- **Version 1.0 (5/14/2014):** Go live
- **Version 2.0 (03/11/2015):** Post-op inclusion and exclusion criteria updated
Medicine is an ever-changing science. As new research and clinical experience broaden our knowledge, changes in treatment and drug therapy are required.

The authors have checked with sources believed to be reliable in their efforts to provide information that is complete and generally in accord with the standards accepted at the time of publication.

However, in view of the possibility of human error or changes in medical sciences, neither the authors nor Seattle Children’s Healthcare System nor any other party who has been involved in the preparation or publication of this work warrants that the information contained herein is in every respect accurate or complete, and they are not responsible for any errors or omissions or for the results obtained from the use of such information.

Readers should confirm the information contained herein with other sources and are encouraged to consult with their health care provider before making any health care decision.
Studies were identified by searching electronic databases using search strategies developed and executed by a medical librarian, Susan Klawansky. Scout and secondary searches were performed in August and November 2013 in the following databases — on the Ovid platform: Medline and Cochrane Database of Systematic Reviews; elsewhere: Embase, Clinical Evidence, National Guideline Clearinghouse and TRIP. For the Scout search retrieval was limited to 1992-current, English language and ages 0-18. In Medline and Embase, appropriate Medical Subject Headings (MeSH) and Emtree headings were used respectively. The search strategy was adapted for other databases as appropriate. The secondary searches on approximately 8 clinical questions were limited to 2003 forward and all infants from birth-23 months. All retrieval was further limited to certain evidence categories, such as relevant publication types, index terms for study types and other similar limits.

May 1, 2014

Flow diagram adapted from Moher D et al. BMJ 2009;339:bmj.b2535


