

Assessment Points				
System	Effect	Assessment by Hx	PE	Test
CV	Extensive microvascular leak, interstitial myocarditis	Rash, swelling	Rash, edema, arrhythmias	ECG, CXR, lytes, BP
RESP	Noncardiac pulm edema, interstitial pneumonitis	Reduced exercise tolerance, dyspnea, cough	Rales by auscultation	CXR, spirometry
GI	Gastroenteritis; liver, spleen, and pancreatic microvascular hemorrhage and edema	N/V, abdominal pain, diarrhea	Abdominal tenderness Hepatosplenomegaly	SGOT, bilirubin
HEME	Thrombocytopenia, anemia	Easy bleeding, malaise	Rash	Hct/Hgb, plts/PT, PTT, BUN
RENAL	Microvascular hemorrhage and edema, interstitial nephritis, prerenal azotemia	Lumbar pain		Cr, lytes
CNS	Meningoencephalitis	Focal defects, deafness, confusion, meningismus, photophobia, seizures		CSF: Checking WBCs and protein
MS	Microvascular hemorrhage, edema	Myalgia, arthralgia	Reduced ROM	

**Key References:** Walker DH, Blanton LS: *Rickettsia rickettsia* and other spotted fever group rickettsiae (Rocky Mountain spotted fever and other spotted fevers). In Bennett JE, Dolin R, Blaser MJ, editors: *Mandell, Douglas, and Bennett's principles and practice of infectious diseases*, ed 8, Philadelphia, PA, 2014, Elsevier, pp 188, 2198–2205.e4; Walker DH: Rocky Mountain spotted fever: a disease in need of microbiological concern, *Clin Microbiol Rev* 2(3):227–240, 1989.

### Perioperative Implications

#### Preoperative Preparation

- Antibiotic therapy and correction of underlying organ system dysfunction
- Surgery only for emergency
- Assess volume, respiratory, and renal status

#### Monitoring

- Consider PA cath, arterial line, UO.
- Intraop ABGs and lytes.
- Platelets and other coagulation variables.

#### Airway

- Severe edema of oropharynx and increasing bleeding tendency can lead to difficult intubation.

#### Induction

- Hypovolemia can cause hypotension.

- Microvascular leak in lung can cause rapid desaturation.
- Increased cardiac arrhythmias.

#### Maintenance

- Owing to CV instability, volume status is key.
- Possibility of respiratory failure and constant volume resuscitation should be anticipated in selecting an anesthetic technique.

#### Extubation

- Oropharyngeal edema and increased bleeding tendency may make reintubation very difficult.

#### Adjuvants

- Vasoactive drugs used in acute resuscitation should be readily available.
- Lidocaine for treatment of cardiac arrhythmias.

#### Postoperative Period

- Intravascular volume shifts; coagulation defects, respiratory failure, CV instability, renal failure

#### Anticipated Problems/Concerns

- Owing to the possibility of multisystem failure, prolonged postop management in the ICU may be required.
- Because early treatment with antibiotics is curative and highly successful in preventing complications, a high index of suspicion (e.g., after tick exposure in endemic areas) is needed.

## Rubella and Congenital Rubella Syndrome

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### Risk

- A rubella epidemic in USA in 1964–1965 resulted in 12.5 million cases of rubella infection and about 20,000 newborns with CRS.
- The number of reported cases of rubella in USA remains low, with a median of 11 cases annually in 2005–2011 because of vaccination.
- The overall burden of CRS is still high in developing countries. There were 66 cases reported in Bangladesh, 26 in Romania, 16 in Nepal, 10 in Zambia, 9 in Japan, and 4 in Sri Lanka in 2014.
- Incidence of cardiac defects in CRS with eye involvement could be as high as 95%. Most common cardiac anomaly in CRS is PDA.

### Perioperative Risks

- CRS is a constellation of multisystem abnormalities
- Such pts may require cardiac surgery like congenital cardiac septal defect correction and/or other non-cardiac surgery like cleft lip/cleft palate repair and a variety of eye procedures under anesthesia. Cataract extraction is an urgent vision saving procedure, so complete optimization (correction of cardiac defects, adequate weight gain) of neonate may not be possible.

### Worry About

- Unexpected difficult intubation in various upper airway anomalies like subglottic stenosis, shortened trachea, and short glottis carinal length associated with many congenital syndromes.

- Hypothermia.
- Hypoglycemia: Exogenous sodium, water and glucose should be provided periop, as they have low GFR and are more prone for hypoglycemia.
- Balance of PVR and SVR.
- Drug metabolism may be deranged because of associated liver abnormalities and hypothyroidism.
- IE prophylaxis is essential, as the turbulent flow produced by the high velocity systolic jet in pulm artery stenosis increases the potential for development of endocarditis.

### Overview

- Rubella is a viral illness characterized by a mild, maculopapular rash. The rubella rash occurs in 50–80% of rubella-infected persons and is sometimes misdiagnosed as measles or scarlet fever.
- Rubella is contagious disease which spreads in droplets. The respiratory secretion, cataractous lens is one of the most infectious materials hence warrants universal precaution.
- CRS is an illness resulting from rubella virus infection during pregnancy. When rubella infection occurs during early pregnancy, serious consequences—such as miscarriages, stillbirths, and a constellation of severe birth defects in infants—can result. The risk of congenital infection and defects is highest during the first 12 wk of gestation and decreases after the 12th week of gestation, with defects rare after the 20th wk of gestation.

- Common congenital defects of CRS include cataracts; congenital heart disease, including PDA, coarctation of aorta, VSD, ASD, and pulm artery stenosis; hearing impairment and developmental delay; brain damage (microcephaly, mental retardation, meningoencephalitis); hepatosplenomegaly; thrombocytopenia; and neonatal jaundice. Other manifestations are type I diabetes mellitus, growth retardation, transient hemolytic anemia, metaphyseal “celery stalking” changes in long bones, transient pneumonitis, transient generalized lymphadenopathy, cryptorchidism, inguinal hernia, and dermal erythropoiesis (“blueberry muffin syndrome”). Infants with CRS usually present with more than one sign or symptom consistent with congenital rubella infection. However, infants may present with a single defect. Hearing impairment is the most common single defect.

### Etiology

- Rubella is a viral illness caused by a togavirus of the genus *Rubivirus*, which is most closely related to group A arboviruses, such as eastern and western equine encephalitis viruses.
- It is an enveloped RNA virus with a single antigenic type that does not cross-react with other members of the togavirus group.
- Rubella virus is relatively unstable and is inactivated by lipid solvents, trypsin, formalin, ultraviolet light, low pH, heat, and amantadine.

- Rubella is transmitted through direct or droplet contact from nasopharyngeal secretions and has an average incubation period of 17 d (range: 12–23 d). Persons with rubella are most infectious when rash is erupting, but they can shed the virus from 7 d before to 7 d after rash onset. CRS develops in an infant as a result of maternal infection in first trimester and

subsequent fetal infection with rubella virus (German measles). Because infants can shed the virus for prolonged periods (up to 1 y of age or longer), infants with CRS should be considered infectious until they are at least 1 y old or until two cultures of clinical specimens obtained 1 mo apart after the infant is >3 mo of age are negative for rubella virus.

### Usual Treatment

- Treatment is supportive. No specific antiviral agent for rubella is currently available.
- Corrective surgeries are typical for other congenital defects.

### Assessment Points

System	Effects	Assessment by Hx	PE	Test
HEENT	Cleft lip/cleft palate Various upper airway anomalies like subglottic stenosis, shortened trachea, and short glottis carinal length	Difficult feeding and poor nutrition	Difficult intubation	Airway assessment
CV	Congenital heart disease, including PDA, coarctation of aorta, VSD, ASD, and pulm artery stenosis; severe PHT	Dyspnea Cyanosis	Tachypnea Rales, rhonchi, wheezing Cyanosis Delayed milestones	ECG CXR 2D ECHO with color Doppler
RESP	Transient pneumonitis	Cough Dyspnea	Poor ventilation, cyanosis Cough, rales, rhonchi, wheezing	CXR
CNS	Brain damage Microcephaly, meningoencephalitis Sensorineural hearing loss	Mental retardation Hearing impairment	Mental retardation Behavioral and language disturbances Hearing impairment	
GI	Associated liver abnormalities Cryptorchidism, inguinal hernia	Neonatal jaundice Hepatosplenomegaly	Deranged drug metabolism	US LFTs
ENDO	Hypothyroidism Type I diabetes mellitus	Growth retardation	Deranged drug metabolism Growth retardation	Thyroid function tests Blood sugar
EYE	Microphthalmia, pigmentary retinopathy, congenital cataract	Affected vision		Ophthalmoscopy
MS	Long bones infection		Metaphyseal “celery stalk” changes in long bones	X-ray long bones
DERM	Blueberry muffin syndrome	Dermal erythroipoiesis	Widespread maculopapular lesions of reddish-blue or magenta color Transient hemolytic anemia, thrombocytopenic purpura, transient generalized lymphadenopathy	Skin biopsy Hematologic investigations

**Key References:** Hariharan U, Garg R, Nagpal VK, et al.: Combined cardiac and noncardiac surgery in an infant with congenital rubella syndrome: an anesthetic challenge, *Paediatr Anaesth* 21(11):1168–1169, 2011; Gaur P, Harde M, Gujjar P, et al.: Unique anaesthesia problems encountered in congenital rubella syndrome, *Int J Adv Case Rep* 2(11):686–688, 2015.

### Perioperative Implications

- Children with CRS require thorough evaluation of associated systemic abnormalities and great vigilance in the periop period.
- Anesthetic goals for pulm artery stenosis include maintenance of a normal or slightly low heart rate, augmentation of preload, and avoidance of factors that increase PVR. For ASD and PDA, maintenance of heart rate with increase in the preload and PVR along with decrease SVR reduces the flow across the defect.
- Anesthetic goals in pts with PS, ASD, and PDA are contrary to each other, but when both these defects are present together, a careful balanced technique has to be maintained to ensure hemodynamic stability as well as to ensure tissue needs are adequately met.
- Anesthetic drugs should be cautiously used due to associated liver abnormalities and hypothyroidism.
- Analgesic management should include avoiding drugs having effect on platelets, as these pts are prone to thrombocytopenia.
- De-airing of IV lines is also an important consideration.
- Combined procedures require change of position intraop, and care should be taken during positioning because of the association of bone abnormalities.
- Concerns related to ductal ligation including effects on blood pressure need to be monitored during the surgical procedure. During the ductal ligation, injury to recurrent laryngeal nerve or accidental ligation of the vessels like pulmonary aorta may occur.

### Preoperative Preparation

- Cardiac status should be optimized prior to noncardiac surgery, which may involve performing cardiac surgery.
- Children should be adequately premedicated to allay separation anxiety and crying which can lead to tachycardia and hypertension.
- IE prophylaxis can be taken care of with cefotaxime 50 mg/kg and amikacin 2 mg/kg in 1 dose prior to surgery.
- Defibrillator and emergency drugs are to be kept ready and arrangements to be made for emergency cardiac surgery in view of difficult resuscitation in these children.

### Monitoring

- Routine monitoring.
- Invasive monitoring with pulm artery cath may be required. Central venous cath may not reflect exact left ventricular status due to presence of PS.
- Transesophageal echocardiography can be more useful than CVP monitoring for evaluation of ventricular filling and function.
- An intra-aortic balloon pump may be kept ready for protection against myocardial ischemia.

### Airway

- CRS may cause difficulty in airway maintenance and unexpected difficult intubation.

### Induction

- IV induction with titrated doses of thiopentone to avoid exposure to higher concentrations of halothane, which would depress the myocardium.

- High dose of narcotics should be avoided; may cause postop respiratory depression, which may alter the PVR.
- Adequate amount of thiopentone sodium and fentanyl helps achieve deeper planes of anesthesia to suppress all presser responses and smooth intubation.

### Maintenance

- Nitrous oxide should be avoided due to its adverse effects on pulm circulation. Air and oxygen with sevoflurane may be used, maintaining the adequate depth of anesthesia to decrease PVR. Vecuronium can be used for muscle relaxation and respiratory support is to be given targeting ETCO<sub>2</sub> in range of 35–40 mm Hg with low tidal volume without PEEP.
- Inj milrinone, a potent pulm vasodilator without marked decrease in systemic vasodilatation, can be used as inotrope if required. IV fluids are to be carefully titrated to avoid cardiac overload.

### Extubation

- Extubation is preferably done in deeper planes to avoid acute arterial and pulm Htn.

### Postoperative Period

- Children should be pain free in postop room, with stable hemodynamic monitors.

### Anticipated Problems/Concerns

- Proper understanding of the hemodynamic effects of the previously mentioned congenital heart diseases and vigilance regarding their anesthetic implications enables us to manage such pts successfully.