Pediatric Cardiovascular Surgery

Preoperative Stabilization of Infants With Hypoplastic Left Heart Syndrome Before Stage I Palliation

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Hypoplastic left heart syndrome (HLHS) is a severe form of congenital heart disease that results in single-ventricle physiology. Single-ventricle physiology requires complete intracardiac mixing of pulmonary venous and systemic venous blood that is then supplied to parallel pulmonary and systemic circuits. HLHS persists throughout a patient’s life. Palliative treatment options are available, but the syndrome has no cure. Management of HLHS requires a series of 3 staged surgical palliation procedures. Over time, survival rates for patients with HLHS have increased because of improvements in surgical technique, postoperative management, and perfusion strategies. Despite these improvements, patients with HLHS have substantial morbidity and a decreased life expectancy. For patients who have a modified Blalock-Taussig shunt, transplant-free survival is 61% at 3 years and 60% at 5 years. For patients who have a right ventricle to pulmonary artery shunt, the survival rates are 67% for 3 years and 64% for 5 years.

CE 1.0 hour

This article has been designated for CE contact hour(s). The evaluation tests your knowledge of the following objectives:

1. Identify prenatal and perinatal factors that affect single ventricle management
2. Describe strategies of perioperative stabilization and management
3. Define 3 major causes of desaturation in single ventricle physiology

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Although a palliative approach for HLHS is widely agreed on in practice, variation occurs in both surgical and medical management. In this article, we review current understanding and practice of preoperative management for infants with HLHS, including perinatal factors that affect management, postnatal stabilization of the infant’s clinical status, and management strategies for balancing single-ventricle physiology and optimizing systemic oxygen delivery.

Perinatal Factors That Affect Preoperative Management

The preoperative management of neonates with HLHS is markedly affected by the timing of the diagnosis (prenatal or postnatal) and perinatal factors, such as the HLHS subtype and the infant’s birth weight, gestational age, and genetic abnormalities. These factors generally contribute to the stability of the infant’s clinical status, whether or not the patient is in extremis, and overall survival.

Prenatal diagnosis of patients with HLHS allows better planning of perinatal care and is strongly associated with superior preoperative clinical status. Studies have indicated that although prenatal diagnosis may not have a marked effect on mortality, it is related to an improvement in morbidity, specifically with regard to neurodevelopmental outcomes. Neonates in whom HLHS is not diagnosed prenatally are more likely to have shock, more severe preoperative lactic acidosis, and a need for inotropic support, which further increases the risk for right ventricular dysfunction and tricuspid regurgitation, both of which are important risk factors for survival of stage 1 Norwood palliation.

Prenatal diagnosis has clear advantages, such as providing time for counseling and educating the infant’s parents and allowing a delivery plan with immediate initiation of prostaglandins and prompt transport to an appropriate care setting. Additionally, prenatal diagnosis of HLHS allows further and more precise delineation of the anatomy, and conditions such as an intact or restrictive atrial septum can be monitored so that plans can be made for prompt balloon atrial septostomy immediately after delivery as needed. Finally, prenatal diagnosis offers the opportunity for fetal interventions such as balloon dilation of the aortic valve or atrial septostomy, which may improve outcomes.

During the perinatal period, delineating the anatomical details and clarifying the HLHS subtype are crucial. HLHS can be due to mitral atresia with aortic atresia, mitral stenosis with aortic stenosis, or mitral stenosis with aortic atresia. The specific subtype of HLHS markedly affects surgical planning (Norwood procedure vs hybrid procedure with stenting of the patent ductus arteriosus and placement of pulmonary artery bands) and may necessitate heart transplant. Some investigators have reported a diminished survival rate of only 79% for patients with mitral stenosis with aortic atresia compared with the other subtypes for stage I Norwood palliation. Furthermore, recent studies have indicated that HLHS with mitral stenosis and aortic atresia is associated with ventriculocoronary connections (sinusoids), which adversely affect outcomes and survival. HLHS with an intact or highly restrictive atrial septum is associated with extremely poor outcomes, with a survival rate of only 68% to 70% before discharge from the hospital after the Norwood procedure for patients deemed to be surgical candidates. However, fetal intervention can increase the survival of this subgroup from 38% to 69%. Furthermore, the Single Ventricle Reconstruction trial indicated that anatomical factors such as the degree of atroventricular valve regurgitation and ascending aortic diameter are associated with increased morbidity and mortality.

Perinatal factors, such as low birth weight, younger gestational age, and genetic abnormalities play a substantial role in an infant’s predicted course and were associated with an increased risk of morbidity and mortality in the Single Ventricle Reconstruction trial. In a retrospective cohort study, Costello et al evaluated neonates admitted to Boston Children’s Hospital, Boston, Massachusetts, during the period 2002 to 2009 who had critical congenital heart disease and a known gestational age. The results indicated that gestational age of 37 to 38 weeks, compared with gestational age of 39 to 40 weeks, was associated with more than 2-fold greater adjusted odds of mortality during hospitalization and
significantly greater morbidity. More recently, Costello et al18 analyzed information in the Society of Thoracic Surgeons Congenital Heart Surgery Database to determine the relationship between gestational age and outcomes for neonates undergoing cardiac surgery. The authors focused on neonates born at early term and concluded that birth during the early term period of 37 to 38 weeks’ gestation was associated with worse outcomes after neonatal surgery. This finding suggests the importance of avoiding early delivery when possible.

**Balancing Circulations to Optimize Systemic Circulation**

Preoperative management of patients with HLHS depends on balancing parallel circulations, which entails maintaining adequate but not excessive pulmonary blood flow while ensuring optimal systemic perfusion. However, the key to management of HLHS patients is optimal systemic perfusion, which can occur in patients with excessive pulmonary blood flow so long as total cardiac output is sufficient. Ineffectively balanced parallel circulation may result in impairment of oxygen delivery, which increases the risk for ischemic injury and contributes to marked infant morbidity and mortality.19

Desaturation in single-ventricle physiology can be attributed to 1 of 3 overall causes: diminished pulmonary blood flow; a low mixed venous oxygen saturation due to a high ratio of total pulmonary blood flow to total systemic blood flow (Qp:Qs ratio), low overall cardiac output, or reduced hemoglobin level; or pulmonary venous desaturation. Astute nursing assessment and ongoing monitoring are necessary to detect these conditions, because the management strategies used are based on the underlying physiology.

Diminished pulmonary blood flow before surgery is a rare occurrence. Generally, even before neonatal pulmonary vascular resistance (PVR) begins to decrease, the PVR is lower than the systemic vascular resistance (SVR) and patients are prone to pulmonary overcirculation. Of note, patients with hypoxia may not have poor blood pressure or metabolic acidosis because of adequate, although hypoxic, systemic tissue perfusion. The primary cause of diminished pulmonary blood flow is an intact or highly restrictive atrial septum; thus, the atrial septum must be evaluated first and ruled out as the cause. Infants with an intact or highly restrictive atrial septum do not respond to oxygen and require prompt evaluation via echocardiography. Early intervention is necessary, with a balloon atrial septostomy, but patients may require venoarterial extracorporeal membrane oxygenation to stabilize their condition before transition to the cardiac catheterization laboratory. In infants with a nonrestrictive atrial septum, physiological and anatomical causes of low Qp:Qs ratio before surgery include elevated PVR related to pulmonary infection and obstruction of pulmonary venous egress due to pulmonary venous obstruction. Management of a low Qp:Qs ratio includes inducing systemic vasoconstriction with medications such as vasopressin or phenylephrine; ensuring functional residual capacity with adequate, but not excessive, positive end-expiratory pressure; and manipulating the PVR by using oxygen, inhaled nitric oxide, sodium bicarbonate, or aggressive sedation. Because a low Qp:Qs ratio is an atypical state before surgery, these therapies should be initiated after acquisition of thorough data and monitoring.

Low mixed venous oxygen saturation with an arteriovenous oxygen difference greater than 25% indicates inadequate cardiac output. Clinical signs may include elevated arterial oxygen saturation with poor perfusion, a wide pulse pressure, oliguria, lactic acidosis, pulmonary edema, and high atrial pressure. Additionally, diminished right ventricular function may be evident on echocardiography. Anatomical and physiological causes of a low mixed venous oxygen saturation include high Qp:Qs ratio related to decreasing PVR, elevated SVR, or systemic outflow obstruction, as with aortic arch obstruction or ductal constriction. Additionally, overall low cardiac output related to diminished ventricular function or tricuspid valve regurgitation will result in reduced mixed venous oxygen saturation. Management of a high Qp:Qs ratio is aimed at optimizing total cardiac output, increasing PVR, and decreasing SVR. Systemic vasodilation with milrinone or sodium nitroprusside may be necessary, and agents such as milrinone, epinephrine, or calcium chloride can be used to optimize cardiac output. Additionally, maximizing oxygen delivery in cyanotic patients by keeping the hemoglobin level in the range of 13 to 16 g/dL is beneficial. Furthermore, in patients with left to right shunting, an increased concentration of hemoglobin increases mixed venous and arterial oxygen.
The ultimate goal for HLHS preoperative management is to optimize systemic oxygen delivery.

Finally, pulmonary venous desaturation causes the mixed blood that is ejected from the heart to be further desaturated. This increased desaturation is caused by ventilation-perfusion mismatch as evidenced by desaturation in patients with findings of atelectasis or pulmonary opacification on chest radiographs or dead-space ventilation. Optimal lung recruitment with positive pressure may prevent pulmonary venous desaturation by optimizing pulmonary gas exchange.20,22

**General Aspects of Preoperative Stabilization and Management**

**Monitoring**

The goals of preoperative management in infants with HLHS include delineation of cardiac anatomy and medical stabilization of clinical status, as well as diagnosis of noncardiac abnormalities such as marked neurological injury, additional organ dysfunction, or genetic abnormalities. A complete postnatal echocardiogram is used to define cardiac anatomy (including HLHS subtype) and critical variables, such as ventricular function, status of the patent ductus arteriosus, the presence and severity of tricuspid regurgitation, and size of the interatrial communication. Complete delineation of cardiac anatomy and overall clinical status are used to determine the need for, type of, and timing of surgical intervention. Patients who have HLHS that was not diagnosed prenatally and who are in shock may require measures to stabilize their condition and evaluation for multisystem organ dysfunction before surgical planning.

During the preoperative period, monitoring tissue oxygen delivery and ensuring an adequate balance between oxygen supply and demand is essential. Of note, high oxygen saturation is a reflection of increased pulmonary blood flow, which may occur at the expense of systemic blood flow, causing systemic hypoperfusion. The ultimate goal is to optimize systemic oxygen delivery. Patients with high total cardiac output (\(\dot{Q}_p + \dot{Q}_s\)) can accomplish this goal regardless of the specific \(\dot{Q}_p: \dot{Q}_s\) ratio, but patients with limited total cardiac output due to ventricular dysfunction or atrioventricular valve regurgitation require careful attention to control of the \(\dot{Q}_p: \dot{Q}_s\) ratio, which can be accomplished by pharmacologically augmenting total cardiac output, limiting systemic oxygen consumption, or early surgical intervention. Adequacy of systemic oxygen delivery can be assessed by clinical examination and monitoring the trends in lactate level, mixed venous oxygen saturation, and findings of near-infrared spectroscopy. Monitoring for adequate hemoglobin concentration is also necessary to ensure optimal oxygen delivery. Mixed venous oxygen saturation can be measured to indicate adequacy of total cardiac output. The difference between arterial oxygen saturation and mixed venous oxygen saturation must be evaluated; a difference greater than 25% may indicate low cardiac output. If a patient does not have appropriate intravenous access for measuring mixed venous oxygen saturation, monitoring via near-infrared spectroscopy can also be used as an adjunct to preoperative management.23 This spectroscopic monitoring is a noninvasive way to determine trends in a patient’s mixed venous oxygen saturation, which will diminish as oxygen delivery decreases or demand increases.24 Furthermore, near-infrared spectroscopy can be useful in detecting hypoxic-ischemic conditions.25 Blood lactate levels are also evaluated regularly to ensure that tissue hypoperfusion is not present; lactate levels are expected to increase as tissues are forced to make the change to anaerobic metabolism.

**Vasoactive Infusions and Inotropes**

One key to preoperative management is maintenance of ductal patency with a prostaglandin infusion. Patients with single-ventricle physiology are dependent on a patent ductus arteriosus for either systemic or pulmonary blood flow. Nearly all infants (with or without the syndrome) have physiological closure of the ductus arteriosus by the fourth day of life, but 20% have functional ductal closure during the first day of life, and more than 80% have ductal closure during the second day of life.26 For patients in shock with suspected ductal closure or a restrictive ductus arteriosus, the initial prostaglandin dose is 0.05 to 0.1 μg/kg per minute; once ductal patency is achieved, the infusion can be decreased to an effective dose of 0.01 to 0.02 μg/kg per minute.27 Using the lowest effective prostaglandin dose to maintain ductal patency is important to minimize the most common dose-dependent side effects of the medication. Hypotension requiring volume replacement and respiratory depression requiring mechanical ventilation may occur. In order to
mitigate the respiratory depression and reduce the need for mechanical ventilation, administration of caffeine can be started with a loading dose of 20 mg/kg and then maintenance dosing of 5 to 10 mg/kg daily.28

Vasoactive medications may be necessary for treatment of cardiogenic shock or support of diminished right ventricular function. For patients whose Qp:Qs ratio is elevated and systemic perfusion is compromised, addition of milrinone might be necessary to increase cardiac output and reduce afterload. However, milrinone may also reduce pulmonary vascular resistance and is associated with a potential undesired risk of increasing the Qp:Qs ratio in excess of total cardiac output.19 Additionally, epinephrine can be used to increase contractility to augment total cardiac output. In neonates, who have an immature sarcoplasmic reticulum, a calcium chloride infusion may be beneficial to improve hemodynamic status, because neonates are highly responsive to exogenous calcium.29 Because infants with HLHS often have elevated heart rates due to low cardiac output, calcium chloride may be the preferred choice because it will increase contractility without further provoking tachycardia. However, monitoring blood levels of calcium and ionized calcium is necessary to avoid supranormal levels. The dose of the vasoactive drug used should be titrated to increase total cardiac output and favorably modify the balance of blood flow between the pulmonary and systemic circulations.

Mechanical Ventilation

Respiratory support may be necessary because of respiratory insufficiency or failure, as well as for low systemic output to reduce oxygen demand. Stabilizing the clinical status of infants with excessive pulmonary blood flow contributing to inadequate systemic blood flow and oxygen delivery before surgery can be a challenge. Previously, mechanical ventilation was used for excessive tachypnea and high oxygen saturation, but in current practice, mechanical ventilation is reserved for patients with respiratory insufficiency or failure or low systemic output.30,31 Spontaneous breathing is preferable, but if necessary, noninvasive ventilatory support can be used to reduce work of breathing and oxygen consumption.

When additional respiratory support is necessary, multiple strategies can be used to increase PVR and thus reduce pulmonary blood flow. Supplemental oxygen decreases PVR and leads to pulmonary overcirculation; thus, avoiding supplemental oxygen is a simple strategy to prevent excessive pulmonary blood flow. Historically, inspired carbon dioxide (to induce hypercarbia) in patients receiving mechanical ventilation and inspired nitrogen (to induce hypoxia) in both spontaneously breathing patients and those receiving mechanical ventilation have been used to increase PVR. For inspired nitrogen, the prevailing thought is that the decrease in oxygen saturation is predominantly due to pulmonary venous desaturation. Although both hypoxia and hypercarbia can decrease the Qp:Qs ratio, only hypercarbia increases cardiac output.30,31 In current practice, permissive hypercapnea is used and is well tolerated, but inspired carbon dioxide is not used. Finally, noninvasive positive pressure ventilation or invasive positive end-expiratory pressure can be used to achieve lung volumes that exceed functional residual capacity. At higher lung volumes, the pulmonary vasculature is compressed, thus increasing PVR and diminishing pulmonary blood flow.20 Positive pressure ventilation decreases afterload on the ventricle by decreasing transmural wall stress. Therefore, intubating a patient has 2 benefits: it decreases oxygen consumption by reducing the work of breathing and decreases afterload, which subsequently increases cardiac output. Patients can benefit from these effects if intubation is necessary because of respiratory failure or low systemic output, but spontaneous breathing remains preferable in the absence of these conditions.

Nutrition

Preoperative enteral feeding in neonates with single-ventricle physiology is controversial and varied. Preoperative enteral nutrition has historically been avoided in patients with single-ventricle physiology and ductal-dependent systemic blood flow because of variable SVR, decreasing PVR, and the potential for systemic hypoperfusion.19 Central venous access may be obtained to provide total parenteral nutrition for adequate caloric intake and volume administration. In general, these neonates require volumes 10% to 20% higher than do healthy neonates to account for the capillary leak that occurs with prostaglandin therapy and additional calories because of the elevated cardiac work imposed by single-ventricle physiology.19 However, clinicians speculate that preoperative enteral feeding may improve

Preoperative enteral feedings may improve nutritional status and thus improve outcomes.
nutritional status and thus improve outcomes, such as decreased length of hospital stay and better neurodevelopmental outcomes. Recent findings indicated that compared with no feeding, preoperative trophic feedings before Norwood palliation were safe and were associated with shorter duration of mechanical ventilation, more stable postoperative hemodynamics, less fluid overload, and earlier postoperative feeding tolerance.

According to a recent survey of medical providers in the United States and Europe, routine preoperative feeding is prescribed by 56% of US providers and 93% of non-US providers. Two-thirds of the respondents who were willing to enterally feed a prostaglandin-dependent neonate did not base their decision on the direction of ductal flow. The majority of providers who preoperatively feed patients who have ductal-dependent lesions offer the infants either breast milk or formula via oral feedings, and some providers prescribe a maximum intake. Feeding tolerance in prostaglandin-dependent neonates given feedings was monitored by using clinical assessment, arterial blood gas analysis, measurement of diastolic blood pressure, and measurement of serum levels of lactate. Some clinicians empirically obtained abdominal radiographs. Intestinal hypoperfusion was one of the primary concerns for withholding enteral feedings, but the relationship of enteral feeding to preoperative necrotizing enterocolitis is unclear.

The incidence of necrotizing enterocolitis is far greater in patients with congenital heart disease than in the general population. Infants with congenital heart disease who are receiving high-dose prostaglandin therapy or who have experienced shock or low cardiac output are at highest risk. In a study on the role of enteral feedings in the development of necrotizing enterocolitis in infants with congenital heart disease, Iannucci et al found that the disease developed in 45 infants (3% of infants studied), 8 (18%) of whom had not undergone cardiac surgery. Obviously the state of the science of preoperative feeding for patients with ductal-dependent single-ventricle physiology is evolving, and thus if enteral nutrition is provided, monitoring is required to evaluate systemic perfusion and signs of feeding intolerance, such as abdominal distention and bloody stools.

**Nursing Considerations**

The goal of preoperative HLHS care is optimizing oxygen delivery and ensuring adequate total cardiac output.

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<tr>
<th>Issue</th>
<th>Strategy</th>
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<tr>
<td>Low pulmonary blood flow</td>
<td>Echocardiogram to assess atrial septum</td>
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<td></td>
<td>May require atrial septostomy</td>
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<td>Systemic vasoconstriction</td>
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<td></td>
<td>Vasopressin, phenylephrine</td>
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<td></td>
<td>Optimize hemoglobin level</td>
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<td></td>
<td>Ensure functional residual capacity with adequate positive end-expiratory</td>
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<td>pressure</td>
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<td>Low mixed venous saturation</td>
<td>Manipulation of pulmonary vascular resistance</td>
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<td>Oxygen</td>
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<td>Inhaled nitric oxide</td>
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<td>Low pulmonary venous</td>
<td>Sodium bicarbonate</td>
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<td>saturation</td>
<td>Aggressive sedation</td>
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<td>Address underlying anatomical and physiological causes</td>
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<td>Optimize total cardiac output</td>
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<td>Hypercarbia</td>
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<td>Systemic vasodilatation</td>
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<td>Milrinone</td>
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<td>Sodium nitroprusside</td>
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<td>Inotropic support</td>
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<td>Calcium chloride</td>
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<td>Maximize hemoglobin level</td>
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<td>Lung recruitment</td>
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<td>Positive end-expiratory pressure</td>
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<td>Consequences of ventilatory maneuvers must be assessed cautiously by</td>
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<td>determining the ventilatory effect on the arterial and mixed venous</td>
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<td>saturation and global end-organ perfusion</td>
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Assessment and monitoring of oxygen saturation, peripheral perfusion, urine output, and feeding intake will inform a nurse if a patient begins to have signs of low cardiac output syndrome. Indications of the syndrome include decreased interest in feedings or feeding intolerance, poor peripheral perfusion, oliguria, and ongoing metabolic acidosis. Additionally, a difference between arterial oxygen saturation and mixed venous oxygen saturation greater than 25% may also indicate low cardiac output. Low cardiac output syndrome is difficult to distinguish from a high Qp:Qs ratio because patients with high arterial oxygen saturation may have adequate systemic output, or may have diminished systemic output as a result of increased pulmonary blood flow occurring at the expense of systemic blood flow. Therefore, astute assessment by nurses and effective communication with the medical providers about subtle clinical changes are crucial. See the Table for management strategies for low pulmonary blood flow, low mixed venous saturation, and low pulmonary venous saturation.
Conclusion
Preoperative management of infants with HLHS is complex and requires multiple complementary strategies. The management and outcomes of these infants are markedly affected by prenatal diagnosis, HLHS subtype, anatomical factors, birth weight, gestational age, and genetic abnormalities. Preoperative management involves complete evaluation, stabilization of the infant’s hemodynamic status, respiratory support, and optimal nutrition strategies. Continued advancement of these strategies is the key to improving outcomes for this complex population of infants. CCN

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None reported.

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To learn more about pediatric cardiovascular surgery, read “Prenatal Counseling and Care for Single-Ventricle Heart Disease: One Center’s Model for Care” by Lafranchi and Lincoln in Critical Care Nurse, October 2015-35:53-61. Available at www.ccnonline.org.

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